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Understanding the Epidemiology and Progression of Systemic Lupus Erythematosus

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Abstract

Objectives—This review examines the burden and patterns of disease in systemic lupus erythematosus (SLE) and the influence and interactions of gender, ethnicity, age, and psychosocial attributes with respect to disease progression, focusing on issues relevant to clinical practice and research.

Methods—PubMed literature search complemented by review of bibliographies listed in identified articles.

Results—An increased risk among reproductive age women is clearly seen in African Americans in the United States. However, in other populations, a different pattern is generally seen, with the highest age-specific incidence rates occurring in women after age 40 years. The disease is 2 to 4 times more frequent, and more severe, among nonwhite populations around the world and tends to be more severe in men and in pediatric and late-onset lupus. SLE patients now experience a higher than 90% survival rate at 5 years. The less favorable survival experience of ethnic minorities is possibly related to socioeconomic status rather than to ethnicity per se, and adequate social support has been shown to be a protective factor, in general, in SLE patients. Discordance between physician and patient ratings of disease activity may affect quality of care.

Conclusions—Our understanding of ways to improve outcomes in SLE patients could benefit from patient-oriented research focusing on many dimensions of disease burden. Promising research initiatives include the inclusion of community-based patients in longitudinal studies, use of self-assessment tools for rating disease damage and activity, and a focus on self-perceived disease activity and treatment compliance.

Keywords

systemic lupus erythematosus; epidemiology; mortality; disease activity; quality of life; ethnicity; socioeconomic status

Systemic lupus erythematosus (SLE or lupus) is a complex and severe rheumatic disease with exceedingly diverse clinical manifestations. Overall improvements in medical care including the availability of antibiotics, antihypertensives, and renal replacement therapy coupled with the judicious use of glucocorticoid, antimalarial, and immunosuppressive drugs have led to improved survival of SLE patients in the past 50 years (1). Despite the improvements in care, patients often suffer long-term morbidity that can adversely affect their quality of life and their ability to work, resulting in substantial direct and indirect costs.

Ethnicity, a broader construct than is implied by the term “race,” encompasses genetic, geographic, cultural, social, and other characteristics shared within a population (2-4). Not surprisingly, the phenotypic expression of lupus varies between individuals of different ethnic groups. A growing body of research has sought to characterize the influence of socioeconomic (5) factors and ethnicity on the incidence, activity, and progression of the disease. Less attention has been paid to the influence of psychosocial factors on disease progression (Fig. 1). In this article, we review relevant research findings pertaining to these dimensions of SLE, focusing on studies conducted in the past 30 years. We also highlight current data gaps and discuss recommendations for future research.

Methods

This review is based on publications found through searches of the MEDLINE database for relevant articles using the combination search terms of lupus and (epidemiology, incidence, prevalence, mortality, gender, ethnicity, damage, quality of life). References within these selected reports were also reviewed. We included epidemiological studies of incidence or prevalence that spanned the years 1975 to 2000, and mortality studies of cohorts assembled in 1975 or later. The mortality studies were limited to inception or near-inception cohorts (that is, cohorts of patients who entered the study within 5 years of diagnosis).

Results

Incidence and Prevalence of SLE Around the World

Incidence rates of SLE range from approximately 1 to 10 per 100,000 person-years (Table 1) (6-24) and prevalence rates generally range from 20 to 70 per 100,000 (Table 2) (6,8-10, 12-18,21-35). In reviewing the available studies, it is important to note that epidemiological studies of SLE may differ by sampling and recruitment methodologies used. The classification criteria initially developed by the American Rheumatism Association, now the American College of Rheumatology (ACR) in 1971 (36) were revised in 1982 (37) and updated in 1997 (38). Early studies, particularly those conducted in hospital settings, failed to capture cases of mild SLE. Studies relying on self-report of an SLE diagnosis, without review of symptom history or medical records, have reported higher prevalence rates (39,40). Another issue with respect to studies that rely on the medical system to ascertain patients is the potential contribution of undiagnosed disease to the total burden within a population. A community survey in Birmingham (United Kingdom), combined with antinuclear antibody testing and clinical assessment of “positive” respondents, reported a prevalence of diagnosed SLE in women ages 18 to 65 years of 54 per 100,000; with the addition of the cases found during the screening, this estimate rose to 200 per 100,000 (41).

A recent study from Olmstead County, Minnesota analyzed SLE incidence rates in 2 periods (6). The age- and sex-adjusted incidence rate was higher in the latter period (1.5 and 5.6 per 100,000 person-years, respectively, in 1950-1979 and 1980-1992). Similar increases were seen in an incidence study in 1980 to 1984, 1985 to 1989, and 1990 to 1994 in Denmark (14). Although some of this increase is likely to reflect an actual increase in disease occurrence, a more accurate ascertainment of cases and the inclusion of milder cases of SLE because of the

wider availability and use of antinuclear antibody testing are also likely to have contributed to these changes. There are few members of ethnic minority groups in these study populations; thus, it is difficult to generalize from these results to the experience in other groups or locations.

The strongest risk factor for SLE is clearly gender: SLE is much more frequent among women than men. In most studies, 90% or more of patients are women, and thus as expected, in studies in which gender-specific rates are given, the incidence and prevalence rates for men are approximately 1/10th those in women. Rates for women and for the total population are shown in Table 1. The rates for the total population are essentially a weighted average of the rates for men and the rates for women. Thus if the rates are 10 times higher in women compared with men, and if there is approximately an equal proportion of males and females in the general population, a weighted average of these rates (ie, the rates for the total population) will be approximately 50% lower than the rates in women. As an example, consider a population in which the incidence rate in men is 1 per 100,000 person-years, and the incidence rate in women is 10 per 100,000 person-years. The average of these 2 rates (assuming equal proportions of men and women in the population) is 5.5 per 100,000 person-years, or approximately half the rate seen in women. This pattern can be seen in Table 1, with total population rates ranging from about 30% to 70% lower than the rates in women.

SLE has been described in 6 continents (Europe, North America, South America, Africa, Asia, and Australia). The disease is rare in Africa but common in African descendants around the world; however, some degree of under-ascertainment is likely to occur in Africa per se (42). Incidence and prevalence rates in people of African or of Asian background are approximately 2 to 3 times higher than in white populations (7,16,17,26,29,43). The disease is also more common among Aboriginal than non-Aboriginal Australians (33,34), and in some First Nations or Native American groups in Canada (9) and the United States (27).

Childhood incidence and prevalence rates of SLE are considerably lower than adult rates. The annual incidence rate of SLE in children (<16 years) was less than 1 per 100,000 persons in studies from Europe and North America (reviewed in (44)). In Taiwan, the prevalence of childhood SLE was estimated as 6.3 per 100,000 (45).

SLE is often described as a disease that most often strikes reproductive-age women. This pattern is clearly seen in data for black women in the United States (Fig. 2). The studies shown in Figure 2 (7,43) also clearly show the increased risk seen among African Americans compared with whites. However, in other populations, a different pattern is generally seen among women, with the highest age-specific incidence rates after age 40 years, as seen in a large study from the United Kingdom (20) as well as in smaller studies from the Sweden and Iceland (12,21) (Fig. 3). Although there are no incident data for Hispanics in the United States or Latin America, the published data suggest they also develop lupus earlier in life (46,47).

Mortality Risk in SLE Patients

Considered in years past a fatal disorder, patients with SLE now live years if not decades after diagnosis. The 5-year survival rate among 99 patients seen at Johns Hopkins University from 1949 to 1953 was 50% (48). In contrast, since the mid-1970s, most studies in Europe, the United States, Canada, and Latin America have demonstrated 5-year survival rates among newly diagnosed patients of over 90%, and 15 to 20 years survival rates of around 80% (Table 3) (5,6,8-10,^{12,21},24,47,49-58). Identification of milder cases, or cases earlier in the disease process, made possible by improvements in diagnostic testing, may contribute to the observed improvements in survival.

Because of the relatively high mortality rate in the first few years after disease diagnosis (59, 60), survival rates are likely to be lower in inception cohorts than in noninception cohorts (ie,

survivor cohorts or left-censored data). Improved survival over time has been noted in analyses of period-specific survival rates since 1980 within adult patients in Canada (61), California (49), Minnesota (6), Sweden (62), and pediatric patients in Taiwan (55). Although the improvement in survival in SLE has been greater than that observed in the general population (63), life expectancy in SLE patients is still below that of comparable demographic groups (6,64,65). Of note, the improvement in survival over time has become evident when examining all causes of death together, but not when examining those deaths attributed to cardiovascular disease (62). It is also important to note that contemporary survival rates in many parts of Asia have yet to reach those of Europe and North America (Table 3), suggesting the importance of economic development and socioeconomic factors in the prognosis of SLE.

Because only about 10% of SLE patients are male, relatively large studies are needed to characterize potential gender-related differences in mortality risk. Most large (>150 patients) inception (49,52-54) and noninception (66-69) cohort studies have reported a lower survival rate among men compared with women; an exception is the study by Reveille and coworkers (70), in which mortality risks for men and women were similar.

Virtually all studies that examine mortality rates for specific ethnic groups (9,49-51,70-73) reported higher mortality risks among the black, Hispanic, and First Nations groups compared with white (or majority) populations. This differential risk by ethnicity was not seen, however, in several of the studies that adjusted for socioeconomic status (eg, by considering income or insurance status or education level) (49,51,71,72,74). This is an important consideration in the interpretation of the role of ethnicity in mortality, given that ethnicity reflects socioeconomic variables and psychological factors that could directly affect mortality risk and that may be amenable to interventions.

Clinical Expression and Progression of Disease

Results pertaining to differences in clinical expression of SLE between men and women are somewhat variable, which may reflect the imprecision often seen in the small samples of men that are included in many of these studies. Larger studies have indicated an increased renal involvement in men compared with women (69,75-79). In the LUMINA (Lupus in Minorities, Nature versus Nurture) study, men accrued damage early in their disease course, predisposing them to accrue more damage subsequently (78,80).

Minority patients by and large have more abrupt disease onset, more severe clinical manifestations, and higher degrees of disease activity compared with white SLE patients (80, 81). Regardless of their age and gender, Hispanic, African American, and Asian SLE patients tend to have more hematological, serosal, neurological, and renal manifestations (29,46,47, 75,82-84). These groups also accrue more damage over time (85) and at a faster pace (86) than white SLE patients and develop specific damage items more often (renal and integument) (85,87,88). Differences observed among ethnic groups early in the course of the disease probably reflect the genetic component of ethnicity, while differences observed later in its course may be more indicative of the nongenetic component of ethnicity such as socioeconomic status and access to medical care (2-5,88).

Pediatric lupus (before age 16 years) often presents with major organ system involvement including renal (46,89-92) and neuropsychiatric (55,92-94) disease. Furthermore, the transition from pediatric to adult care may be quite difficult for these patients. Independence and responsibility are highly desirable but they should be balanced with adequate social support and the discussion of issues particularly pertinent to this age group such as safe sexual practices and the prevention of other risky behaviors. Late-onset lupus (age 50 years and above) tends to have a more insidious onset (95,96), less major organ system involvement (97), and lower degrees of disease activity (98,99), yet these patients tend to have a poor outcome, in terms of

both damage accrual (85,100) and mortality (51,96,101,102). Other factors associated with age (comorbidities) probably explain these findings (103).

The development of clinical instruments for the assessment of disease activity (104) and disease damage (105) has facilitated research on SLE progression. Recent studies have reported differences in the way patients and physicians perceive the impact of SLE. In terms of disease activity, for example, patients tend to score subjective manifestations, which overall indicate how well they feel, whereas physicians tend to score objective manifestations, particularly laboratory parameters (106-108). Disease activity scores using visual analog scales are often discordant between patients and physicians, with 28% differing by ± 2.5 cm in 1 study (107), and 58% differing by ± 1.0 cm in another study (106). This difference may have implications for disease management. For example, if a physician scores higher than the patient, some level of noncompliance is likely to occur; in the opposite situation, patients may request treatment beyond what physicians think is necessary. In terms of damage, patients may also put more weight on manifestations that impact on their appearance, body image, or self-esteem (eg, extensive skin scarring) than in features that are less tangible or symptomatic (proteinuria or diminished renal function, for example) (109).

Within the social context, social support has been identified as a modulating factor in disease activity, damage accrual, and even functioning (80,85,110-114). Karlson and coworkers, for example, in a cross-sectional study of 200 SLE patients from 5 centers, found that lack of social support was associated with poorer physical and mental functioning (114), whereas social support was negatively associated with disease activity in the LUMINA study (80). There may be important variation in these effects among ethnic groups, however. Social support was associated with better mental health outcomes in black and in white SLE patients in the 5-center study described previously, but some of the other positive effects of social support were only seen among whites or patients in the higher socioeconomic status groups (42). In addition, lower levels of social support and higher degrees of helplessness and of abnormal coping styles have been consistently shown in Texan-Hispanics and to a lesser extent in African Americans, than in Caucasians and Puerto Rican-Hispanics from the LUMINA cohort. Patients from these minority ethnic groups tend, overall, to experience worse disease outcomes (5,46). Adequate social support acts as a protective factor or buffer, making it possible for patients and their families to navigate the health and social systems, utilize them, and benefit from them. This is the case not only in patients with SLE but also in patients with other chronic diseases (115-118). Moreover, interventions aimed at improving social support have been shown to have some benefits, *albeit* modest, in SLE (119). Other socioeconomic parameters such as education, marital status, occupation, and income contribute to the overall social context in which the disease occurs and progresses.

Acculturation is the exchange of cultural features when two cultures come in contact with each other for the first time. Changes in both cultures may occur but the 2 cultures remain distinct. In the case of Hispanics moving into the United States, a higher degree of acculturation indicates that more features of the American culture had been acquired. The term is applied principally to the minority culture (immigrants) adopting features of the majority culture (host country). Discrimination and acculturation are two aspects of the social context that have been less well explored as to their impact on disease activity, damage, and functioning. These two constructs have been explored in the LUMINA patients. Neither inadequate acculturation by Texan-Hispanic patients into the mainstream American culture nor self-perceived discrimination by patients regardless of their ethnic category were found to have a direct impact on disease activity, damage accrual, or the patients' self-reported levels of functioning (118, 120,121).

Health-Related Quality of Life

The level of self-reported physical and mental functioning or self-reported health-related quality of life is another important dimension to consider in SLE. Most studies have used the 36-item short form (SF-36) of the Medical Outcomes Survey (122) and have shown that patients with SLE function at levels below those of the general population (123-126). Health-related quality of life appears to be mediated not only by specific disease manifestations, by the degree of disease activity and of damage accrued, but also by the patient's level of helplessness and ability to cope with the disease. Two recent studies using shorter forms of this instrument (127,128) reported a significant reduction of overall self-perceived levels of well-being in LUMINA patients (129), or a reduction in physical function score in the Carolina Lupus Study patients compared with a general population comparison group (52). In the LUMINA study, these measures were affected not only by disease-related parameters but by social support, helplessness, and abnormal illness-related behaviors (129).

Discussion

We have reviewed the published literature related to the incidence and prevalence rates of SLE as well as the factors affecting the expression and the intermediate and long-term outcomes of the disease, including mortality risk, organ damage, and health-related quality of life. The disease is overall more frequent among minority population groups around the world and the outcome of the disease is less favorable in these populations. The role of socioeconomic factors in the course and outcome of the disease is noteworthy, and given the association of poor socioeconomic and minority status, the role of both in the final outcome of the disease may be difficult to disentangle. SLE at the extremes of life (pediatric and late-onset) may be particularly severe.

Significant advances have been made in the past 2 decades in understanding the genetic predisposition to disease occurrence and the underlying pathophysiological mechanisms accounting for the inflammatory process and the irreversible organ damage that may ensue over time. In addition, methodological developments have enabled a standardized ascertainment of disease activity, damage, function, and quality of life in the patient affected with lupus. Basic research continues to play an invaluable role in understanding the disease and its course, and this knowledge will hopefully translate to better therapeutic options for those affected with SLE. Despite these advances, significant gaps in our knowledge remain, and other important areas of research need to be addressed by those working in the field.

Several specific recommendations can be made based on our assessment of limitations and gaps in our understanding of the epidemiology and progression of SLE. For example, little research exists pertaining to the incidence or prevalence of SLE in many areas (particularly outside of Europe and North America) or among some groups within the United States (eg, Hispanics and Asian Americans). Data from countries that are undergoing a transition to industrial economies would be useful in determining the impact of these changes on disease risk. The different age patterns in incidence rate among women from different ethnic, socioeconomic, or geographic areas raise important questions for clinicians and researchers (eg, does the higher rates in older women in some areas reflect an increased diagnosis of milder disease?).

Research designs that allow for the separate examination of socioeconomic factors and ethnicity in studies of disease severity and progression could provide insights that would lead to the development of more focused and productive interventions. In terms of disease progression, studies based on the previously constituted cohorts may not be inclusive enough of the less severe cases of lupus managed by community rather than by academic rheumatologists. It is important to design studies that include community-based patients in

longitudinal observational studies. Studies from disease registries may be useful for incidence or prevalence data if a specific population base or catchment can be defined and, can be useful for study of long-term outcomes if systematic follow-up is achieved. One issue with respect to mortality studies is the source of data relating to SLE-related mortality. Reliance on cause of death data obtained from death certificates is likely to lead to significant under-ascertainment of the impact of SLE (130). This is likely to be less of an issue in clinic-based cohort studies that are actively following patients than in population studies relying on death certificate to ascertain SLE-related mortality.

The development of self-assessment tools for the ascertainment of disease activity (131) and damage (132) may help facilitate these study designs. It is also important to determine the impact of lupus in other important outcomes not assessed with currently available instruments. Generic instruments such as the SF-36 or the SF-6D do not address specific domains that are quite relevant for those affected with the disease. Some of the outcomes that warrant more attention include body image and the perception of self-esteem. In addition, the relationship between self-perceived disease activity and compliance with treatment needs to be further explored. As part of the education and empowering process, lupus patients need to understand the “hidden” manifestations of the disease and appreciate their possible detrimental consequences, so compliance with treatment regimens will improve.

The agenda for the next generation of lupus researchers, while quite vast, promises to improve our understanding of the disease. Coupled with laboratory research, clinical- and population-based research will translate into better outcomes for patients afflicted with this disease.

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Figure 1.

A model of the factors affecting the course and outcome of SLE (modified from (5)).

Figure 2.

Age-specific incidence rates for SLE in females, from 2 studies from the United States with data for African Americans and whites. (A) is from Hochberg (43) and (B) is from McCarty and coworkers (7). (Color version of figure is available online.)

Figure 3.

Age-specific incidence rates for SLE in females in the United Kingdom (20) (calculated by interpolation from graph), Sweden (12), and Iceland (21). (Color version of figure is available online.)

Table 1

Incidence of Systemic Lupus Erythematosus in Adults, by Location, in Studies Spanning 1975 to 2000

Author (Reference), Country (Area), Study Period ^a	Total Rate per 100,000 per year ^b (n)	Female Rate per 100,000 per year ^b (n)
Americas		
Uramoto (6), United States (MN), 1980 to 1992	5.6 (48)	9.4 (42)
McCarty (7), United States (PA), 1985 to 1990	2.4 (191)	
	African Americans	9.2 (45)
	Whites	3.5 (129)
Naleway (8), United States (WI), 1991 to 2001	5.1 (44)	8.2 (36)
Peschken (9), Canada (Manitoba), 1980 to 1996	First Nations ~ 3.5 (49)	—
	Whites ~ 1.2 (177)	
Nossent (10), Curaçao 1980 to 1989	Afro-Caribbean 4.6 (68)	7.9 (60)
Vilar (11), Brazil, 2000	8.7 (43)	14.1 (38)
Europe		
Stahl-Hallengren (12,13), Sweden, 1981 to 1991	1981-86 4.5 (38)	1981-86 5.4 (32)
	1987-91 4.5 (41)	—
Voss (14), Denmark, 1980 to 1994 ^c	1980-84 1.0 (—)	—
	1985-89 1.1 (—)	—
	1990-94 2.5 (—)	—
Nossent (15), Norway, 1978 to 1996	2.9 (83)	5.1 (73)
Johnson (16), United Kingdom (Birmingham), 1991	Total 3.8 (33)	Total 6.8 (31)
	—	Afro-Caribbean 22.8 (6)
	—	Asian 29.2 (8)
	—	Whites 4.5 (17)
Hopkinson (17,18), United Kingdom (Nottingham), 1989 to 1990	Total 4.0 (23)	Total 6.5 (19)
	Afro-Caribbean 31.9 (3)	—
	Whites 3.4 (19)	—
Nightingale (19), United Kingdom, 1992 to 1998	3.0 (390)	5.3 (349)
Somers (20), United Kingdom, 1990 to 1999	4.7 (1638)	7.9 (1374)
Gudmundsson (21), Iceland, 1975 to 1984	3.3 (76)	5.8 (67)
López (22), Spain, 1998 to 2002	2.2 (116)	3.6 (102)
Alamanos (23), Greece, 1982 to 2001	1.9 (178)	—
Oceania		

Author (Reference), Country (Area), Study Period ^a	Total Rate per 100,000 per year ^b (n)	Female Rate per 100,000 per year ^b (n)
Anstey (24), Australia, 1986 to 1990	Aboriginal 11 (13)	—

Abbreviations: MN = Minnesota, PA = Pennsylvania, WI = Wisconsin.

^a Nightingale (19) and Somers (20) used the United Kingdom General Practitioner Research Database; Gudmundsson (21) used hospital records, and all other studies used various type medical records for case ascertainment.

^b Age-adjusted rates provided when available; group specific estimates provided when based on 2 or more cases. —, data not reported.

^c Voss (14) included a total of 107 patients, but the number per time period was not provided.

Table 2

Prevalence of Systemic Lupus Erythematosus in Adults, by Location, in Studies Spanning 1975 to 2000

Author (Reference), Country (Area), Study Period ^a	Total Rate per 100,000 ^b (n)	Female Rate per 100,000 ^b (n)
Americas		
Uramoto (6), United States (MN), 1992	130 (—)	—
Maskarinec (25), United States (HI), 1989	Total 42 (454)	Total 74 (401)
		Non-whites 78 (315)
		Whites 71 (86)
Chakravarty (26), United States (CA, PA), 2000	California 108 (—)	Total 184 (—)
		African American 406 (—)
		Hispanic 139 (—)
		Asian, Pacific Island 93 (—)
		Whites 164 (—)
	Pennsylvania 150 (—)	Total 253 (—)
		African American 694
		Hispanic 245 (—)
		Asian, Pacific Island 103 (—)
		Whites 203 (—)
Naleway (8), United States (WI), 2001	79 (64)	132 (54)
Boyer (27), United States (AK), 1991	112 (9)	166 (8)
Peschken (9), Canada (Manitoba), 1996	Total 22 (257)	—
	First Nations 42 (49)	—
	Whites 21 (177)	—
Nossent (10), Curaçao, 1989	Afro-Caribbean 48 (69)	84 (63)
Europe		
Stahl-Hallengren (12,13), Sweden, 1986, 1991	1986 42 (44)	—
	1991 68 (41)	—
Voss (14), Denmark, 1994	22 (104)	38 (93)
Nossent (15), Norway, 1995	50 (89)	89 (79)
Hochberg (28), United Kingdom, 1982	7 (20)	13 (20)
Johnson (16), United Kingdom (Birmingham), 1991	Total 28 (242)	Total 50 (227)
	Afro-Caribbean 112 (50)	Afro-Caribbean 197 (48)
	Asian (Indian) 47 (36)	Asian (Indian) 97 (35)
	Whites 21 (155)	Whites 36 (143)
Hopkinson (17,18), United Kingdom (Nottingham), 1990	Total 25 (147)	Total 45 (136)
	Afro-Caribbean 207 (21)	—
	Asian (Indian) 49 (7)	—
	Asian (Chinese) 93 (2)	—
	Whites 20 (117)	—

Author (Reference), Country (Area), Study Period ^a	Total Rate per 100,000 ^b (n)	Female Rate per 100,000 ^b (n)
Samanta (29), 1989, United Kingdom (Leicester)	Total 26 (50)	Total -
	Asian 64 (19)	Asian (Indian) 73 (13)
	Whites 20 (31)	Whites 32 (26)
Molokhia (30), United Kingdom (London), ages 15 to 64, 1999	—	Afro Caribbean 177 (72)
	—	West African 110 (20)
	—	Whites 35 (66)
Gourley (31), Ireland, 1993	25 (415)	—
Gudmundsson (21), Iceland, 1984	36 (86)	62 (77)
Lopez (22), Spain, 2002	34 (367)	58 (324)
Alamanos (23), Greece, 2001	38 (193)	67 (170)
Middle East		
Al-Arfaj (32), Saudi Arabia, 1992	19 (2)	37 (2)
Oceania		
Bossingham (33), Australia, 1996 to 1998	Total 45 (108)	—
	Aboriginal 93 (26)	—
Segasothy (34), Australia, 1999	Aboriginal 74 (18)	—
	Whites 19 (6)	—
Anstey (24), Australia, 1991	Aboriginal 52 (13)	Aboriginal 100 (13)
Hart (35), New Zealand, 1980	Total 18 (136)	—
	Polynesians 51 (34)	—
	Whites 15 (96)	—

Abbreviations: AK = Alaska, CA = California, HI = Hawaii, MN = Minnesota, PA = Pennsylvania, WI = Wisconsin.

^aChakravarty (26) used state hospitalization databases, adjusted for estimate of proportion of SLE patients hospitalized annually. Hochberg (28) used the United Kingdom General Practitioner Research Database. Gudmundsson (21) used hospital records. Al-Arfaj (32) used a survey with follow-up examination, and all other studies used various type medical records for case ascertainment.

^bAge-adjusted rates provided when available; group-specific estimates provided when based on 2 or more cases. —, data not reported.

Survival Probabilities for Systemic Lupus Erythematosus Patients in Cohorts of Newly Diagnosed or Recently Diagnosed Patients Constituted Since 1975

Table 3

Author (Reference), Country (Area), Study Period ^a	Survival % by years Follow-up					
	N Total	N Deaths	5	10	15	20
Americas						
Pistiner (49), United States (CA), 1980 to 1989	195	—	97	93	—	—
Kasitanon (50), United States (MD), 1987 to 2004	1378	118	95	91	85	78
African-Americans	543	69	—	—	—	71
Whites	767	47	—	—	—	84
Uramoto (6) United States (MN), 1980 to 1992	48	—	95	72	—	—
Naleway (8), United States (WI), 1991 to 2001	44	8	88	76	—	—
Alarcón (5,51) United States (AL, TX), 1994 to 2006						
African-American	221	32	90	—	—	—
Hispanic	117	17	87	—	—	—
Whites	176	13	94	—	—	—
Campbell (52), United States (NC, SC), 1995 to 2001	265	32	90	—	—	—
Peschken (9), Canada (Manitoba), 1980 to 1996						
First Nations	49	—	95	82	75	—
Whites	177	—	98	95	92	—
Nossent (132) Curaçao, 1980 to 1990	68	25	60	46	—	—
Pons-Estel (47), Latin America, 1997 to 2003 ^b	1214	34	95	—	—	—
Europe						
Gudmundsson (21), Iceland, 1975 to 1988	76	17	84	78	—	—
Stahl-Hallengren (12), Sweden, 1981 to 1996	81	17	93	83	—	—
Alamanos (53), Greece, 1981 to 2001	185	21	96	87	—	—
Manger (54), Germany, 1985 to 1999	338	35	97	90	—	—
Asia						
Wang (55), Taiwan (pediatric), 1980 to 1990	52	32	60	44	—	—
1991 to 2001	101	18	85	78	—	—
Mok (56), Hong Kong, 1992 to 1999	182	9	93	—	—	—

Author (Reference), Country (Area), Study Period ^a	N Total	N Deaths	Survival % by years Follow-up			
			5	10	15	20
Kasitanon (57), Thailand, 1986 to 2000 ^b	349	52	84	75	—	—
Murali (58), India, 1981 to 1993	98	23	77	60	—	—
Oceania						
Anstey (24), Australia, 1984 to 1991	22	9	60	—	—	—

Abbreviations: AL = Alabama, MN = Minnesota, NC = North Carolina, PA = Pennsylvania, SC = South Carolina, TX = Texas, WI = Wisconsin.

^a Group-specific estimates provided when based on 2 or more deaths. Data for Kasitanon (50) (survival rates for total population), Uramoto (6), and Peschken (9) estimated from graphs. —, data not reported. Alarcón (51) included patients diagnosed up to 5 years before study entry; median time from diagnosis to study was 18 months. Campbell (52) included patients diagnosed up to 4 years before study entry; median time from diagnosis to study was 13 months. Kasitanon (50) accounted for difference between diagnosis and study entry using delay entry into the risk sets in calculating survival in hazards models. All other studies were inception cohorts (entering all patients at time of diagnosis).

^b Pons-Estel (47) presented 4-year survival probability.