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## The Incidence and Prevalence of Systemic Lupus Erythematosus, 2002–2004: The Georgia Lupus Registry

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### Abstract

**Objective**—The Georgia Lupus Registry is a population-based registry designed to improve our ability to estimate incidence and prevalence of systemic lupus erythematosus (SLE) in a large population.

**Methods**—Potential cases were identified from multiple sources during the years 2002 through 2004. Cases were defined by the American College of Rheumatology (ACR) Criteria for SLE or a combined definition. Age-standardized rates were determined and stratified by race and sex. With capture-recapture analyses, we estimated the under-ascertainment of cases.

**Results**—Using the ACR case definition, the overall crude and age-adjusted incidence rate was 5.6/100,000, with capture-recapture and combined definition rates being slightly higher. The age-adjusted incidence rate for women was >5 times higher (9.2 vs. 1.8) than that for men. Black women had an incidence rate nearly 3 times higher than that for white women with a significantly higher rate in the 30 to 59 years age group. The overall crude and age-adjusted prevalence rates were 74.4 and 73/100,000, respectively. The age-adjusted prevalence rate for women was nearly 9 times higher (127.6 vs. 14.7) than that for men. Black women had very high rates (196.2). A striking difference was seen in the proportion with end-stage renal disease in prevalent cases, with a sevenfold greater involvement among blacks.

**Conclusion**—With more complete case finding, our incidence and prevalence rates are among the highest reported in the United States. Results continue to underscore striking gender, age, and racial disparities between blacks and whites.

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In the 1950's, systemic lupus erythematosus (SLE) was thought to be rare, predominantly afflicting females with light hair, fair skin, and "inability to tan" (1). An epidemiologic study from 1956–65 showed for the first time the higher burden of disease in black women compared to their white counterparts (2). We now appreciate the disproportionate burden of SLE on women, particularly in their childbearing years, and in certain racial groups. These epidemiologic studies advanced our understanding of the burden of SLE but were limited in their ability to find all cases in the population and thus describe the full spectrum of diagnosed SLE.

Given the recent significant increase in awareness of and research in SLE, along with the availability of innovative techniques (3), the purpose of this Georgia Lupus Registry (GLR) study is to advance our epidemiologic understanding of SLE by doing more complete case finding in a targeted population, avoiding referral bias in a particular institution, using available case definitions to better define the incidence and prevalence of diagnosed SLE, and characterizing individuals with this disease from a population perspective. The GLR is one of two recently completed Centers for Disease Control and Prevention (CDC) funded population-based lupus registries designed to minimize many of the limitations of previous studies. An innovative tool in this approach is the use of the state public health surveillance exemption to the Health Insurance Portability and Accountability Act (HIPAA) to acquire greater access to protected health information without requiring individual patient consent, a limitation that can bias findings. This novel and powerful approach allows for an unprecedented completeness of case finding from multiple sites of ascertainment throughout the targeted community. Coupled with detailed training of abstractors, strict quality control of data gathering and processing, multiple sources of case ascertainment that minimizes bias from a consent process or institution type, and the high number of cases, this study provides more reliable population-based estimates of incidence and prevalence of SLE than previously reported.

## Patients and Methods

### THE GEORGIA LUPUS REGISTRY

In 2002, the CDC Arthritis Program funded the Georgia Department of Public Health (GA DPH) to conduct surveillance of SLE in 2 Georgia counties with large black populations (Fulton and DeKalb) (3). To avoid biased ascertainment and underreporting as a result of recruiting large numbers of community patients, the GA DPH, as a "public health authority", used its public health surveillance exemption to the HIPAA Privacy Rule (45 CFR parts 160 and 164) to obtain protected health information (PHI) without written patient consent (45 CFR 164.512[b]). PHI was needed to determine if diagnosed cases met the various case definition criteria and to provide enough information to prevent duplicate counting of patients when the same patient was encountered in multiple facilities. The GA DPH contracted with Emory University as its designated agent to provide lupus expertise and manage the project. CDC considered this surveillance project to be "public health practice" (rather than research) that did not need CDC Institutional Review Board (IRB) review, but it was reviewed and approved by the IRB's at Emory University and the GA DPH.

## STUDY POPULATION AND PERIOD

The study population consisted of residents of Fulton and DeKalb counties, which include the city of Atlanta. Prevalence was estimated for 2002 and incidence for 2002–2004. The Bureau of the Census estimate in 2002 for the two counties was 1,552,970 with 51.1% women, 49.3% blacks, and 46.4% whites (4) and remained stable in 2003–04. The registry captured diagnosed cases retrospectively. Case ascertainment efforts began in late 2004 in order for data elements to have had time to be captured in various records and data systems. Given the tremendous effort required to obtain and validate cases through multiple sources in the pluralistic US health care system, data collection was not completed until 2011.

## CASE DEFINITIONS

SLE is a complex disease that is currently defined using a variety of case definitions. The gold standard case definition is diagnosis by expert clinical assessment, usually a rheumatologist, which is impractical for large population-based studies. We used 3 case definitions for this study:

1. 4 ACR criteria. The most widely accepted standard is meeting 4 of 11 criteria in the 1997 update of the 1982 American College of Rheumatology (ACR) Revised Classification Criteria for SLE (4 ACR criteria) (5, 6).
2. Treating rheumatologist's diagnosis. Those with 3 ACR criteria were required to have a documented statement of diagnosis of SLE in the medical record by a board-certified rheumatologist. This acknowledges that a clinical diagnosis by the treating rheumatologist with direct access to the patient is important, particularly in prevalent patients with longstanding disease where certain records may have been inaccessible or lost over time for our study.
3. <4 ACR criteria plus lupus kidney disease. In the absence of fulfilling 4 of 11 ACR criteria, those with SLE renal involvement were defined by either:
  - a. a biopsy consistent with class II–VI lupus nephritis (7–9). These biopsies were linked to SLE administrative coding or clinical documentation and were deemed significant enough of a clinicopathologic finding to suggest a case has SLE in the absence of meeting full ACR criteria (10).
  - b. end-stage renal disease (ESRD) requiring dialysis or renal transplantation with documentation of SLE in the medical record. This avoids missing patients who may have spent most of their time at dialysis centers for care and/or were busy dealing with other comorbidities. The likelihood of being able to locate or access medical records where most ACR criteria are documented decreases for these patients. Incident and prevalent patients may have achieved ESRD status before 2004 or 2002, respectively.

For analytic purposes, we report results in two ways: for the standard case definition (4 ACR Criteria) alone and for a “combined” case definition that includes all 3 case definitions.

Incident cases were defined as those newly diagnosed with SLE from January 1, 2002 through December 31, 2004 and prevalent cases were defined as those with a diagnosis of

SLE of any duration during the year 2002. Both must also have met a case definition with a documented address in one of the two targeted counties during the time of interest.

### **CASE ASCERTAINMENT, SCREENING, AND VALIDATION**

The primary sources of potential cases included hospitals, rheumatologists, nephrology groups, and dermatology groups in and around the catchment area (Figure 1).

Administrative databases were queried retrospectively for the International Classification of Diseases, Ninth Revision, Clinical Modification (ICD-9-CM) diagnostic code 710.0 (SLE), as well as 695.4 (discoid lupus), 710.8 (other specified connective tissue disease), and 710.9 (unspecified connective tissue disease). Antiphospholipid antibody syndrome, which lacks a specific ICD-9-CM code, was searched for if a consistent code was utilized at a particular facility.

Secondary sources included regional commercial and hospital-based laboratories, which were queried respectively for anti-nuclear antibodies (ANA) titer 1:320, anti-DNA and/or anti-Sm antibodies, depressed complement levels, and anti-cardiolipin antibodies. Pathology laboratories were queried for cutaneous and renal biopsies consistent with lupus. Data were also obtained from the Veterans Administration, electronic medical record systems, and the US Renal Data System (USRDS), which captures all patients with end stage renal disease receiving any kidney replacement therapy (e.g., dialysis or kidney transplantation).

After final screening for residency in the target counties during the target time period, available medical records were abstracted for over 200 data elements, each with detailed definitions in a data dictionary; elements were sought continuously from all sources (without stopping when a certain number of case definition criteria were met). Demographic information, such as race, was obtained from the medical record. Date of diagnosis was the earliest date assigned in the medical record by the patient's treating physician because the date at which the patient met 4 ACR criteria, often used to define incidence in prospective studies, was difficult to accurately ascertain retrospectively.

Abstractors were thoroughly trained and tested by board-certified rheumatologists who were expert in lupus before entering the field, where they continued to undergo quality assessments every 100 records. This required a minimum inter-observer agreement of 90% of all elements and 95% of ACR criteria using the Principal Investigators' (SL, CD) abstraction as the gold standard or remedial training until those criteria were met.

### **STATISTICAL ANALYSIS**

Crude SLE incidence rates and 95% confidence intervals as well as race- and sex-stratified rates were estimated using methods based on the Poisson distribution (11). Denominator data for estimating rates for DeKalb and Fulton counties for the years 2002–2004 were obtained from the postcensal population estimates (4). Age-adjusted estimates and 95% confidence intervals were calculated based on the standard 2000 projected age distribution by the direct method using R (routine ageadjust.direct) (12), which calculates age standardized (adjusted) rates and “exact” confidence intervals based on the gamma distribution (13). Similar methods were used to estimate 2002 prevalence.

## CAPTURE-RECAPTURE

Capture-recapture (C-RC) methods estimate the completeness of case ascertainment when using multiple information sources. Log-linear models were used to estimate the true SLE population size by evaluating the degree of overlap among three data sources – hospitals, specialists, and health care systems, which were chosen by expert opinion from the investigators (SL, CD) to be the primary sources of cases. Modeling was performed separately for incident and prevalent data based on those patients who met the case definition of 4 ACR Criteria. The log-linear model was used to estimate the number of persons who were missed in the population.

Seven hierarchical log-linear models were fit to the data (one model assuming independence among the three data sources; three models of pairwise interaction; and three models of two pairwise interactions). The best fitting model was determined by goodness of fit statistics and the parsimony principle. Based on the estimated undercount, revised (C-RC) estimates of incidence and prevalence were calculated. All (C-RC) analyses followed the methods described by Bishop et al and McCarty et al and were implemented using SAS Proc Genmod (14, 15).

## Results

### INCIDENCE - 4 ACR CRITERIA

In 2002–2004, 267 cases fulfilled 4 ACR criteria for SLE, including 196 blacks, 62 whites, and 9 other races (Table 1). The overall crude and age-adjusted incidence rates were similar [5.6 per 100,000 person-years]. Age-adjusted incidence rates were 5 times higher among women than men [9.2 (95% CI, 8.1–10.5) vs. 1.8 (95% CI, 1.3–2.4)]. Among women, age-adjusted rates for blacks were nearly 3 times higher than that for whites (13.4 vs. 4.7); among men, rates for blacks were >4 times higher than that for whites (3.2 vs. 0.7). C-RC analysis estimated 31 (95% CI, 18–55) missed SLE cases resulting in a higher C-RC adjusted rate of 6.3 per 100,000 person-years.

Overall age-specific rates were significantly different between blacks and whites in both women and men. Figure 2 shows age-specific rates by sex and race. Black women had significantly higher IR's compared to white women in the 30–59 age range, especially in the 30–39 age strata (Figure 2A). Age-specific differences were not found between blacks and whites in men, though there were significantly fewer numbers of men in these strata (Figure 2B).

The overall average age at diagnosis was 40.5 (s.d.±16.5) years, with no significant difference between women and men (40.7±16.3 vs. 39.4±17.9 years, p=0.48). However, blacks were significantly younger at diagnosis compared with whites (39.4±15.9 vs. 45.4±17.7 years, p=0.016). Among women, the only incident cases in those <12 years of age were blacks. In the 12–19 age strata, black women and men predominated, with white cases only coming from women.

### INCIDENCE – COMBINED CASE DEFINITION

The “combined” case definition yielded an additional 78 cases and a total of 345 incident cases, increasing the crude and adjusted rates to 7.3 and 6.9 per 100,000 person-years, respectively (Table 1). Of these 78 cases, 72 met 3 ACR criteria, 13 had a renal biopsy consistent with lupus nephritis (5 of these had a documented anti-nuclear antibody (ANA) and/or anti-DNA antibody), and none had ESRD. All patients were identified as black or white except for 11 Asians and 3 with unknown race.

Using the “combined” case definition, the age-adjusted rate for women was 6 times higher than that for men (11.7 vs. 1.9 per 100,000 person-years). The age-adjusted rate for blacks was 3.2 times higher than that for whites (10.7 vs. 3.3 per 100,000 person-years). The proportions were different when comparing gender by race. The rate in women was 2.9 times higher in blacks than whites (17.0 vs. 5.8 per 100,000 person-years). Black men had a 4.3 times greater rate compared with white men (3.4 vs. 0.8 per 100,000 person-years).

### PREVALENCE - 4 ACR CRITERIA

In 2002, 1,156 cases fulfilled 4 ACR criteria for SLE, including 889 blacks, 251 whites, and 16 other races (Table 2). The overall crude prevalence rate (PR) was 74.4 (95% CI, 70.3–78.9) per 100,000 person-years. Age-adjusted PRs were similar to the crude rates. The age-adjusted PR for women was nearly 9 times higher than that for men (127.6 vs. 14.7). Among women, the PR for blacks was >3 times higher than that for whites (196.2 vs. 59); among men, the PR for blacks was >3 times higher than that for whites (23.7 vs. 7.5). C-RC analysis was done only for the 4 ACR criteria case definition and estimated 133 (95% CI, 98–181) missing SLE cases resulting in a C-RC adjusted PR of 83 per 100,000 person-years.

### PREVALENCE – COMBINED CASE DEFINITION

The “combined” case definition yielded an additional 290 cases for a total of 1,446 prevalent cases, increasing the crude and adjusted PRs to 93.1 and 92.1 per 100,000 person-years, respectively (Table 2). Of these 290 cases, 213 met 3 ACR criteria, 52 had a renal biopsy consistent with lupus nephritis (27 of these had a documented ANA and/or anti-DNA antibody), and 49 had ESRD. Of those with ESRD, 11 had a documented ANA and/or anti-DNA antibody and all had less than 3 ACR criteria (8 had a documented ANA and/or anti-DNA antibody).

The age-adjusted PR for women was >8 times higher than that for men (159.8 vs. 19.6). The age-adjusted PR for blacks was >3 times higher than that for whites, 147.5 vs. 43.1 per 100,000 person-years. When comparing the age-adjusted PRs in women, blacks were >3 times higher than whites (241.5 vs. 77.7); in men, blacks were >3 times higher than whites (32.2 vs. 9.6). Overall rates were significantly different between blacks and whites in both women and men. Black women had significantly higher PRs compared to white women across all age strata except for ages 0–11 and 70 (Figure 2C). For men, PRs were noticeably higher in blacks compared to whites between ages 20 and 49 (Figure 2D).



## CLINICAL MANIFESTATIONS

Among incident cases with 4 ACR criteria, arthritis, hematologic, and serologic ACR Criteria were the most common through the first year after the diagnosis (Table 3). The next most frequent were renal disorder and serositis in blacks, and photosensitivity and oral ulcers in whites. Less than 2% of incident cases had documented ESRD through the first year after the diagnosis. However, among prevalent cases, the proportion of blacks with ESRD was 8.4% and whites 1.2%.

## Discussion

SLE is one of the most challenging conditions to study on a population level. Several studies throughout the world have attempted to advance our epidemiologic knowledge of SLE and results have varied widely. We have limited our review in this manuscript to those in the North American region (Table 4). Discrepancies in rates are in part due to the inherent disparities of SLE (i.e., higher rates in certain ethnic groups). Other reasons include the use of different case definitions, biased sources for case ascertainment, small source populations, the different demographic groups targeted, the protean manifestations of the disease that make diagnosis difficult, the poor reliability of self-report, the lack of reliability in coding in health system databases, and poor access to health care for high-risk populations. These latter differences not only exist across different countries and health care systems, but also within the same country. Although no one study can address these limitations completely, the methodologic advances in this study resulted in some of the most reliable population-based estimates of incidence and prevalence of SLE.

Leveraging the state's public health surveillance exemption to obtain and review patient information from medical records and various databases without consent was a powerful tool to maximize case ascertainment that had never been utilized before for SLE. This led to a better population-based assessment without compromising patient confidentiality. This also required significant effort in maximizing the quality of the data, which was enhanced through the collection of uniformly defined data elements, regular quality assessments of the abstractors and data, and sharing of best practices with a parallel registry in Michigan through monthly teleconferences facilitated by the CDC. Multiple sources of cases captured a wide spectrum of disease, both phenotypically and the degree of disease severity. This included all major sources of pediatric cases. The large numbers of cases led to greater precision of the estimates and power to compare certain groups.

How cases are defined is essential to a study's interpretation and comparability to other studies. The "gold standard" for diagnosing SLE is by clinical assessment from an experienced clinician (i.e., a rheumatologist), which is often impractical for population-based studies. Currently, the most commonly accepted definition is the updated 1997 ACR Criteria for the Classification of SLE (5, 6). While the use of ACR criteria enhances the comparability of research studies, the sensitivity of the 1982 criteria has been shown to be only 83% in an external population versus 96% in the test population. The criteria also tend to be skewed towards limited detection of mild cases of SLE and incident cases at early stages of their prodrome. The population size would, therefore, be underestimated and biased towards those of longer disease duration and greater severity. Epidemiologic studies

would benefit from alternative definitions for comparison. Since the fulfillment of a single case definition in the field did not limit the extent of information that was obtained, this study utilized an alternative definition that improved sensitivity while minimizing impact to specificity, which is acceptable for a large epidemiologic study. Previous studies often utilized one definition. Alternative definitions, if available, were often those who met incomplete ACR criteria. Diagnoses validated with administrative data or by self-report have limitations and should be supplemented with additional data in situations when medical review is not feasible (16, 17).

The burden of SLE in the Atlanta, Georgia area is significant, with an overall age-adjusted prevalence rate among the highest reported in the US at 73 per 100,000 person-years by ACR Criteria, 83 by C-RC, and 92.1 by combined case definition. The overall age-adjusted incidence rate is similarly high at 5.6 per 100,000 person-years by ACR Criteria, 6.3 by C-RC, and 6.9 by combined case definition. Use of the combined case definition yielded higher rates and underscores some limitations of the ACR Classification Criteria, which should be viewed as a minimal estimate in this retrospective study given the potential for data to be missed or overlooked (18). High rates may, in part, be due to improved awareness of SLE leading to increased referrals and testing, as well as improved 5-year survival rates that have gone from <50% to >90% due to earlier diagnosis and more aggressive treatment (19, 20). Including undiagnosed or early cases, which were not addressed in this study, would raise these estimates.

Striking gender, age, and racial disparities in SLE have been confirmed. Women have an age-adjusted incidence rate >5 times higher and prevalence rate >8 times higher than men using the ACR Criteria. The relatively higher female to male ratio that we found in prevalent (8:1) compared to incidence cases (5:1) may be related to a greater awareness among physicians of SLE in general and as a result in men. Likewise, mortality has been reported to be relatively higher in men than in women, particularly by studies from the 1980's and 1990's (21). As the diagnosis and life expectancy of SLE improves, further mortality studies will advance our understanding of potential differences in the burden of disease and its outcomes by sex. Blacks have an incidence and prevalence rate >3 times higher and develop SLE earlier than whites. SLE particularly burdens black women, with some of the highest incidence and prevalence rates ever reported (15, 22). Black women between the ages of 30–59 are at particularly high risk for developing SLE (Figure 2A). In women <20 years old, there were only 4 incident cases in whites as compared to 20 in blacks. Only black women developed SLE in ages <12 (n=5). Although age-specific prevalence rates were significantly higher for black women compared to their white counterparts across all age groups, the difference was more striking between the ages of 30 and 69 (Figure 2C). In men, blacks also had higher prevalence rates compared to whites, particularly between ages 20 and 50 (Figure 2D). Our findings suggest that different age-related patterns of mortality by sex and race may occur in SLE patients as indicated in prior studies (23).

The burden of ESRD continues to be high, afflicting 6.7% and 1.9% of prevalent and incident cases ( 4 ACR criteria), respectively. A striking difference was seen in the proportion with ESRD, where there was no racial difference among incident cases but a 7 fold greater involvement among blacks among prevalent cases. Some of this can be



explained by an inherently higher risk in those of African descent to develop nephritis (24). However, it also indicates a strong potential for disparities in health care access and other socioeconomic factors (25). 49 out of 127 prevalent ESRD cases did not have at least 3 ACR criteria indicating that up to a third of ESRD cases from lupus nephritis may not meet ACR criteria. A consistent renal biopsy, particularly in the setting of suggestive autoantibodies, is felt to be indisputable evidence for SLE and should be considered as sufficient “stand alone” clinical criteria in prospective studies (26). In an epidemiologic study such as this, given that SLE patients with ESRD are seen less often by rheumatologists and nephrologists outside dialysis centers, documentation of many of these serologies may have been lost with archived or destroyed records.

There are several limitations to this study. First, the US healthcare system is complex, heterogeneous, and fragmented, requiring that each practice and institution be approached separately to voluntarily participate in this study. Second, data were collected retrospectively from medical records designed for clinical use and varied tremendously with respect to organization, legibility, and accessibility. The result of these 2 limitations was a labor-intensive process that required work into 2011 to evaluate the study period of 2002–04. Although trained abstractors were audited periodically to ensure consistency and accuracy, a degree of variability of clinical diagnosis by rheumatologists cannot be excluded. As a result of the retrospective nature of this registry, the degree in which the experience of treating physicians impacting the definition of cases, particularly of those with milder disease or fewer number of ACR criteria, cannot be determined. Third, there is underascertainment of serologic tests, particularly in prevalent cases in which diagnostic tests could have occurred many years ago and the results lost in the records, thus leading to lower rates of documented ANA and other autoantibodies than what would have been expected in a prospective study (table 3). Fourth, the catchment area was defined by artificial county boundaries. Fifth, the results of this study are best generalized to whites and blacks in the Southeastern US and not to other racial/ethnic groups and in other regions or countries. Sixth, race/ethnicity was assigned based primarily on the physician assessment as documented in the medical record. This may not reflect the patient’s true self-identity. Finally, this study was not able to estimate the rate of undiagnosed disease.

The GLR advances our epidemiologic understanding of SLE, a complicated and difficult to diagnose disease, and confirms the significant burden of SLE, particularly in younger black women and ESRD in blacks compared to whites. It is part of a larger, coordinated effort to more accurately estimate the burden of SLE among other high-risk groups in the US, such as Hispanics, Asians, and American Indians/Alaska Natives. Experience from this study could also inform efforts to develop ongoing surveillance of SLE and other medical conditions for which similar challenges exist.

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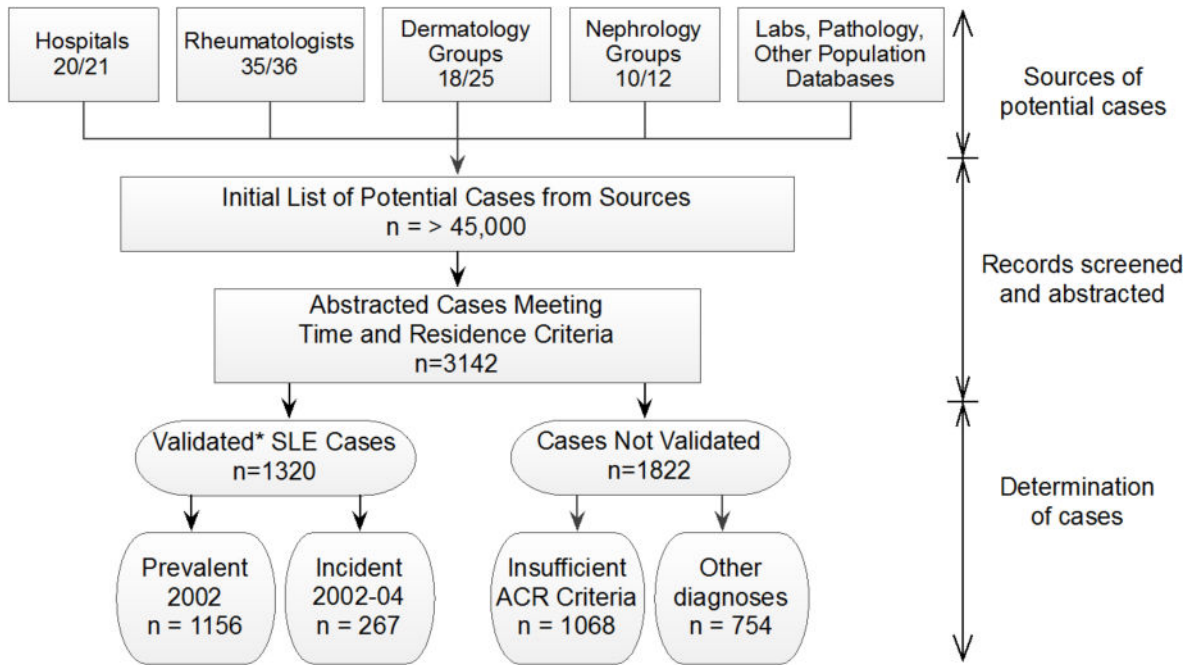
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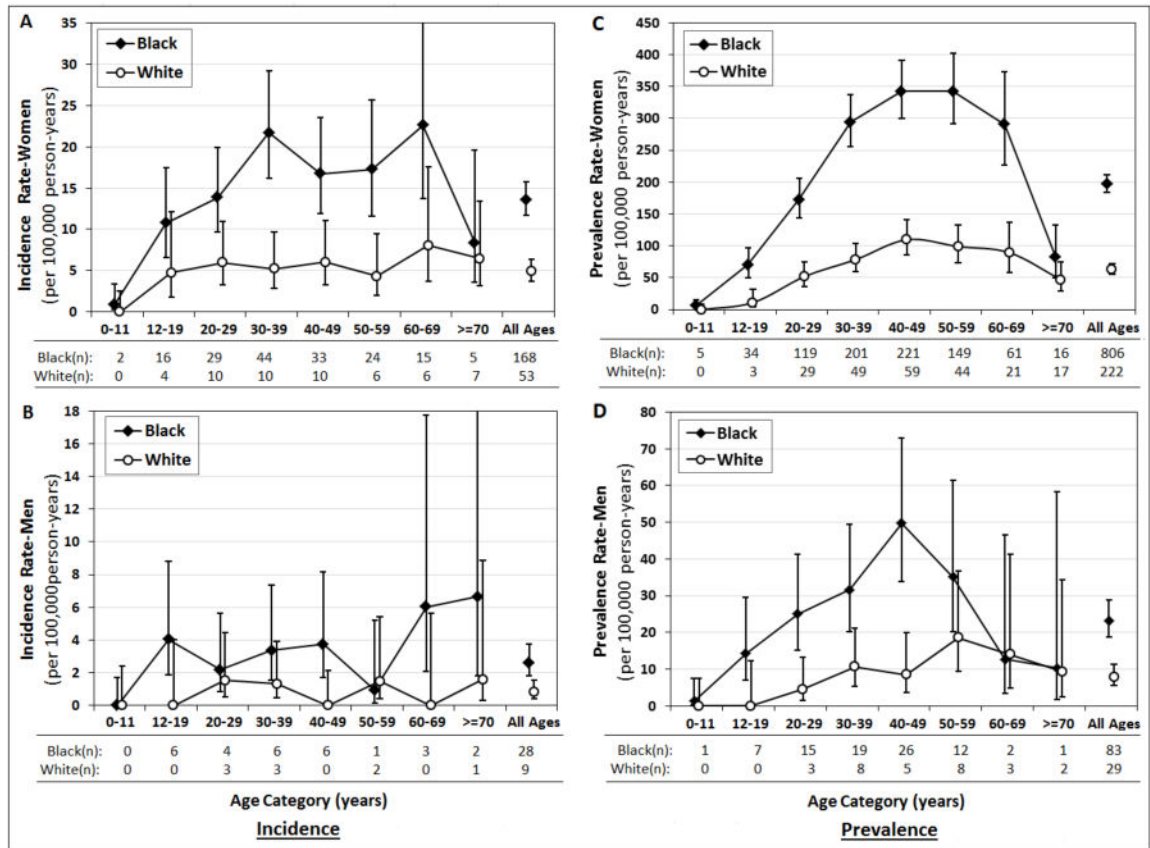
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\* ≥ 4 ACR Criteria; 103 incident cases in 2002 were also counted as prevalent

**Figure 1.** Flow Chart of Case Finding (for ≥ 4 ACR criteria case definition) from Primary and Secondary Data Sources to Screening/Abstracting to Meeting Case Definitions



**Figure 2.**  
Age-Specific Incidence and Prevalence Rates of SLE by Sex and Race ( 4 ACR Criteria Case Definition)



**Table 1**

Incidence of Systemic Lupus Erythematosus (SLE) in Fulton and DeKalb Counties, GA (Jan. 1, 2002–Dec. 31, 2004), Overall and by Race/Sex for 2 analytic case definitions

Race/sex	Catchment area population (person-years)	4 ACR Criteria				Combined Case Definition <sup>a</sup>			
		No. of Cases	Crude Rate (95% CI)	Age-Adjusted Rate <sup>b</sup> (95% CI)	Capture-Recapture		No. of cases	Crude Rate (95% CI)	Age-Adjusted <sup>b</sup> Rate (95% CI)
					Missed <sup>c</sup>	C-RC Rate <sup>d</sup>			
Overall	4,742,264	267	5.6 (5.0,6.3)	5.6 (5.0,6.3)	31 (18,55)	6.3 (5.6,7.0)	345	7.3 (6.5,8.1)	6.9 (6.2,7.7)
women	2,424,592	228	9.4 (8.3,10.7)	9.2 (8.1,10.5)	28 (15,51)	10.6 (9.3,11.9)	300	12.4 (11.1,13.9)	11.7 (10.4,13.1)
men	2,317,672	39	1.7 (1.2,2.3)	1.8 (1.3,2.4)	4 (1,17)	1.9 (1.4,2.5)	45	1.9 (1.5,2.6)	1.9 (1.4,2.6)
Black	2,321,302	196	8.4 (7.3,9.7)	8.7 (7.6,10.0)	22 (11,43)	9.4 (8.2,10.7)	252	10.9 (9.6,12.3)	10.7 (9.5,12.1)
women	1,239,819	168	13.6 (11.7,15.8)	13.4 (11.5,15.6)	20 (10,42)	15.2 (13.1,17.5)	221	17.8 (15.6,20.3)	17.0 (14.9,19.5)
men	1,081,483	28	2.6 (1.8,3.7)	3.2 (2.3,4.5)	3 (1,10)	2.9 (2.0,4.1)	31	2.9 (2.0,4.1)	3.4 (2.5,4.7)
White	2,210,389	62	2.8 (2.2,3.6)	2.7 (2.1,3.5)	8 (2,23)	3.2 (2.5,4.0)	79	3.6 (2.9,4.5)	3.3 (2.7,4.2)
women	1,082,131	53	4.9 (3.7,6.4)	4.7 (3.6,6.2)	5 (1,22)	5.4 (4.1,6.9)	68	6.3 (5.8)	5.8 (4.6,7.5)
men	1,128,258	9	0.8 (0.4,1.5)	0.7 (0.4,1.4)	1 (0,16)	0.9 (0.5,1.6)	11	1.0 (0.5,1.7)	0.8 (0.4,1.6)

Rates are per 100,000 person-years.

<sup>a</sup> [ 4 ACR criteria] or [3 ACR criteria with final diagnosis of SLE by rheumatologist] or [ $\leq$ 4 ACR criteria and (i) biopsy consistent with class II-VI lupus nephritis or (ii) end-stage renal disease secondary to SLE requiring dialysis/renal transplantation]

<sup>b</sup> Age adjusted using the 2000 projected US population

<sup>c</sup> Estimated missing count (95% CI)

<sup>d</sup> Capture-recapture adjusted incidence rate (95% CI)

**Table 2**  
Prevalence of Systemic Lupus Erythematosus (SLE) in Fulton and DeKalb Counties, GA (2002), Overall and by Race/Sex for 2 Case Definitions

Race/sex	Catchment area population	4 ACR Criteria						Combined Case Definition <sup>d</sup>		
		No. of Cases	Crude Rate (95% CI)	Age-Adjusted Rate <sup>b</sup> (95% CI)	Capture-Recapture		No. of Cases	Crude Rate (95% CI)	Age-Adjusted Rate <sup>b</sup> (95% CI)	
					Missed <sup>c</sup>	C-R Rate <sup>d</sup>				
Overall	1,552,970	1,156	74.4 (70.3,78.9)	73.0 (68.9,77.4)	133 (98,181)	83.0 (78.6,87.7)	1,446	93.1 (88.4,98)	92.1 (87.4,97)	
women	794,310	1,043	131.3 (123.6,139.5)	127.6 (120,135.7)	115 (83,159)	145.8 (137.6,154.4)	1,300	163.7 (155,172.8)	159.8 (151.3,168.8)	
men	758,660	113	14.9 (12.4,17.9)	14.7 (12.2,17.6)	20 (8,49)	17.5 (14.8,20.8)	146	19.2 (16.4,22.6)	19.6 (16.7,23.0)	
Black	765,475	889	116.1 (108.8,124.0)	118.5 (111.1,126.5)	91 (64,131)	128.0 (120.3,136.3)	1,094	142.9 (134.7,151.6)	147.5 (139.2,156.4)	
women	408,642	806	197.2 (184.1,211.3)	196.2 (183,210.2)	79 (54,115)	216.6 (202.8,231.3)	985	241.0 (226.5,256.6)	241.5 (226.9,257.1)	
men	356,833	83	23.3 (18.8,28.8)	23.7 (19.1,29.3)	13 (4,42)	26.9 (22,32.8)	109	30.5 (25.3,36.8)	32.2 (26.9,38.7)	
White	720,292	251	34.8 (30.8,39.4)	32.7 (28.8,37.2)	36 (18,73)	39.8 (35.5,44.7)	328	45.5 (40.9,50.7)	43.1 (38.5,48.1)	
women	352,914	222	62.9 (55.2,71.7)	59.0 (51.5,67.5)	33 (15,74)	72.3 (63.9,81.7)	292	82.7 (73.8,92.8)	77.7 (69.0,87.5)	
men	367,378	29	7.9 (5.5,11.3)	7.5 (5.2,10.9)	4 (0,19)	9.0 (6.4,12.6)	36	9.8 (7.1,13.6)	9.6 (6.9,13.4)	

Rates are per 100,000 person-years.

<sup>a</sup> [4 ACR criteria] or [3 ACR criteria with final diagnosis of SLE by rheumatologist] or [ $<4$  ACR criteria and (i) biopsy consistent with class II-VI lupus nephritis or (ii) end-stage renal disease secondary to SLE requiring dialysis/renal transplantation]

<sup>b</sup> Age adjusted using the 2000 projected US population

<sup>c</sup> Estimated missing count (95% CI)

<sup>d</sup> Capture-recapture adjusted prevalence rate (95% CI)

Table 3

11 ACR Criteria Manifestations of SLE plus End-Stage Renal Disease (ESRD) among Incident and Prevalent Cases, by Race

Manifestations	4 ACR Criteria + ESRD									
	Incident <sup>a</sup> , n (%)			Difference (Black-White) (95% CI)			Prevalent <sup>b</sup> , n (%)			
	Overall n=267	Black n=196	White n=62	Black n=1156	Black n=889	White n=251	Difference (Black-White) (95% CI)			
Malar Rash	55 (20.6)	38 (19.4)	12 (19.4)	374 (32.4)	272 (30.6)	93 (37.1)	-6.5 (-17.7, 4.8)			
Discoid Rash	40 (15.0)	32 (16.3)	7 (11.3)	262 (22.7)	225 (25.3)	37 (14.7)	10.6 (-2.2, 23.3)			
Photosensitivity	43 (16.1)	25 (12.8)	17 (27.4)	295 (25.5)	199 (22.4)	92 (36.7)	-14.3 (-25.7, -2.8)			
Oral Ulcers	61 (22.8)	42 (21.4)	18 (29)	250 (21.6)	175 (19.7)	73 (29.1)	-9.4 (-21.4, 2.6)			
Arthritis	167 (62.5)	122 (62.2)	38 (61.3)	779 (67.4)	600 (67.5)	169 (67.3)	0.2 (-7.8, 8.2)			
Serositis	91 (34.1)	74 (37.8)	15 (24.2)	491 (42.5)	386 (43.4)	100 (39.8)	3.6 (-7.2, 14.4)			
Renal	91 (34.1)	75 (38.3)	12 (19.4)	394 (34.1)	326 (36.7)	60 (23.9)	12.8 (0.8, 24.8)			
ESRD <sup>c</sup>	5 (1.9)	4 (2)	1 (1.6)	78 (6.7)	75 (8.4)	3 (1.2)	7.2 (-6.6, 21.1)			
Neurologic	24 (9.0)	18 (9.2)	6 (9.7)	156 (13.5)	126 (14.2)	29 (11.6)	2.6 (-10.5, 15.8)			
Hematologic	216 (80.9)	160 (81.6)	49 (79)	870 (75.3)	672 (75.6)	184 (73.3)	2.3 (-4.9, 9.5)			
Immunologic	187 (70.0)	143 (73)	38 (61.3)	736 (63.7)	584 (65.7)	139 (55.4)	10.3 (1.2, 19.4)			
Antinuclear Antibody	244 (91.4)	180 (91.8)	56 (90.3)	949 (82.1)	738 (83)	199 (79.3)	3.7 (-2.5, 10.0)			

<sup>a</sup>Manifestations through the first year after diagnosis

<sup>b</sup>Manifestations through 12/31/2002

<sup>c</sup>Defined by abstraction and the USRDS database

**Table 4**

Population-based Studies of Incidence and Prevalence of SLE in North America, including the Caribbean and Puerto Rico

Study years	Study location/Country	Population at risk (Race)	Case Definition	Case Ascertainment Sources	Number of SLE cases	Incidence*	Prevalence**
1956-65	New York City and Jefferson County, AL/US (2)	1,165,700 (White and Black)	Clinical suspicion, characteristic serologic and pathologic findings	Hospital files, selected clinics and rheumatologists, LE cell tests	Total White Black	2.0 <sup>^</sup> (overall)	19 <sup>^</sup> (overall)
1965-73	San Francisco, CA/US (27)	121,444 members of Kaiser (All races)	4 1971 ARA criteria by chart review	Outpatient diagnoses from internists and dermatologists	Total	7.6 (overall)	51 (overall)
1970-77	Baltimore, MD/US (28)	905,759 (1970)-786,775 (1980)	4 1971 ARA criteria by chart review	Hospital discharge (SLE)	Total White Black	4.6 <sup>^</sup> (overall)	ND
1980-90	Island of Curacao/Netherlands Antilles (29)	146,500 (~95% African Caribbean, <5% White)	4 1982 ACR criteria by chart review	Hospital discharge records, all internal medicine and dermatology specialists by physician report, death certificates in Public Health Department	Total (1980-90) 90 African Caribbean Prevalent (1990) Incident (1980-89)	4.6 (overall) 7.9 (Females) 1.1 (Males)	48 (overall) 84 (Females) 9 (Males)
1985-90	Allegheny County, PA/US (15)	1,336,449 (All races)	4 ACR criteria validated by chart review	Rheumatologists, hospitals, university database (SLE)	Total White Black Other	2.4 (overall)	ND
1950-92	Rochester, MN/US (30)	29,885 (1950) -70,745 (1990) (94% White)	4 1982 ACR criteria by review of 430 medical records	Community diagnostic retrieval system (SLE, ANA, LE cell, false positive syphilis)	Total	3.1 <sup>+</sup> (overall) 5.6 <sup>+</sup> (1980-92) 1.5 <sup>+</sup> (1950-79)	122 <sup>^</sup> (Jan 1992)
1980-96	Province of Manitoba/Canada (31)	~1,100,000 (NAI and NI)	4 1982 ACR criteria validated by chart review	Regional arthritis center DB and the medical records of all rheumatologists, hematologists, nephrologists, and general internists	Total NAI Non Indian	2.0-7.4 (NAI) 0.9-2.3 (NI)	42.3 (NAI) 20.6 (NI)
1997	Nogales, AZ/US (32)	19,489 (92% Mexican American)	4 1982 ACR revised criteria (definite) or 3 ACR revised criteria (probable) by chart review & exam	Community referrals to lupus evaluation center, practice DB search (SLE)	Total Definite Probable	ND	94 (overall)
1990-99	Martinique Island/French West Indies (33)	381,427 (Mostly African Caribbean)	4 1982 revised ACR criteria by investigator chart review	Hospital electronic records, (university and community hospitals), practitioners and software of specialists, independent practitioners, files	Total	4.7 (overall)	64.2 (overall)

Study years	Study location/Country	Population at risk (Race)	Case Definition	Case Ascertainment Sources	Number of SLE cases	Incidence*	Prevalence**
				of antinuclear antibodies and antiphospholipid, file for Social Security, mortality DB of antinuclear antibodies and antiphospholipid, file for Social Security, mortality DB of antinuclear antibodies and antiphospholipid, file for Social Security, mortality DB			
2000	NHANES III (sample of US population) (34)	20,050 (All races)	Self-reported MD diagnosis +/-SLE drugs	Self-reported physician diagnosis from NHANES III	Self-report On SLE drugs 40 12	ND	241 (self-report) 54 (on SLE drugs)
1991–2001	Rural area North Central Wisconsin/US (35)	77,280 (97% White)	4 1982 ACR revised criteria (definite) or 1–3 ACR criteria (incomplete) by chart review	Community clinic electronic records (SLE)	Total Definite Incomplete 174 117 57	5.1 <sup>^</sup> (definite) 8.2 <sup>^</sup> (definite Females) 1.9 <sup>^</sup> (definite Males)	79 <sup>^</sup> (definite) 132 (definite Females) 25 <sup>^</sup> (definite Males)
2003	Puerto Rico/US (36)	552,733 private insured people (Race ND)	ICD-9 code 710.0	All insurance claims submitted by health care providers (physicians, dentists, laboratories, pharmacies, and hospitals)	Total Females Males 877 812 65	ND	159 (overall) 277 (Females) 25 (Males)
1994–2003	Quebec/Canada (37)	~7.5 million (All races)	2 claims of SLE (>2 months and <2 years apart)	Administrative data: billing codes, hospitalization data and procedure code data	Total (2003) 3825	3.0 (PB) 2.8 (HDD)	33 (PB) 33 (HDD) 51 (Composite) 45 (Bayesian model)
2008–10 depending on region	Mexico City, Nuevo Leon, Yucatan, Sinaloa, Chihuahua/Mexico (38)	Sample of 19,213 people age 18 and above (Race ND)	4 1982 revised ACR criteria by study rheumatologist assessment	COPCORD methodology <sup>a</sup> ; community-based survey directed physician assessments	NA	ND	60 <sup>^</sup> (overall) 80 (Females) 40 (Males) 90 (Mexico City) 40 (N Leon) 70 (Yucatan) 40 (Sinaloa) 40 (Chihuahua)
2000–09	Barbados (39)	268,792 (93% African origin)	4 1982 ACR revised criteria	Clinic charts from public and private rheumatology services, letters and calls to physicians	Total Females Males 183 172 11	6.59 <sup>^</sup> (total) 12.1 <sup>^</sup> (Females) 0.77 <sup>^</sup> (Males)	84.1 (overall) 152.6 (Females) 10.1 (Males)
2002–04	Atlanta, GA/US	1,552,970 (49.3% Black, 46.4% White)	4 1982 revised ACR criteria or “combined” definition by review of charts and labs	Hospitals, rheumatologists, dermatologists, nephrologists, laboratories, pathology labs, VA, USRDS	Total Females Males 1446 1300 146	6.9 <sup>^</sup> (total) 1.7 <sup>^</sup> (Females) 1.9 <sup>^</sup> (Males)	92.1 <sup>^</sup> (total) 159.8 <sup>^</sup> (Females) 19.6 <sup>^</sup> (Males)

\* Per 100,000 per year

\*\* per 100,000

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age adjusted to national or regional population

age and sex-adjusted

COPCORD: Community Oriented Program for the Control of Rheumatic Diseases

Abbreviations: ACR-American College of Rheumatism; NA-not available; ND-not determined; NAI-North American Indian; NI-Non Indian; DB-database; PB-physician billing; HDD-hospital discharge data; GP-general practitioners; MR-medical record; VA-Veteran's Administration; USRDS-US Renal Data System.