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Epidemiology of Systemic Lupus Erythematosus: an update

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Abstract

Purpose of review—Systemic lupus erythematosus (SLE) is the prototypical systemic autoimmune disease with a significant disease burden across the world among different ethnic, racial and age groups. The pathophysiological understanding of SLE is constantly evolving and with it, the need for a better definition of the disease itself, for understanding the risk among the different affected populations, and for identifying the factors responsible for the damage accrual through the years.

Recent findings—More accurate estimates of incidence and prevalence of SLE among different ethnicities and minority groups not only in the USA, but in Europe, Middle East, and Asia have provided new insights into the disease burden around the world. Despite advances in treatment, mortality among SLE patients remains high with significant geographic variations.

Summary—Gender, race, and ethnicity significantly affect SLE incidence, prevalence, damage accrual, and mortality.

Lupus in the USA

In 2014, data from the Michigan (1) and Georgia (2) Lupus Registries were published and provided a valuable insight into the incidence and prevalence of systemic lupus erythematosus (SLE) in the United States in a population predominantly composed of Caucasians and African Americans (Table 1). A similar CDC funded population-based registry (3), determined the prevalence and incidence of SLE among American Indian and Alaska Native people within the Indian Health Service clinical population. These studies overcame the shortcomings of previous epidemiologic data in the USA by using identical case definitions (meeting ≥4 American College of Rheumatology (ACR) criteria or a renal biopsy of lupus nephritis/end-stage renal disease or a rheumatologist's diagnosis), and a broad range of case-finding sources. The number of Hispanic and Asian patients in these three registries was small, so the CDC supported the creation of two similar lupus registries in California and New York. The recently published data from the California Lupus Surveillance Project (CLSP) (4) and the Manhattan Lupus Surveillance Program (MLSP) (5) included populations with greater numbers of Asian and Hispanic patients. Both studies reported a higher prevalence and incidence rate of SLE in women compared with men, and in African Americans compared to Caucasians, similar to the data reported from Michigan and Georgia. The diverse population allowed estimation of incidence and prevalence among

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Hispanics and Asians, who had a higher incidence and prevalence of SLE compared to Caucasians, but lower than African Americans (table 1). The diverse populations included in these two registries allowed the first reliable estimation of incidence and prevalence among the Hispanic and Asian population in the United States.

Ungprasert et al. (6) used both the SLICC and the American College of Rheumatology SLE criteria to investigate the incidence of SLE in Olmsted County, Minnesota, from 1993 to 2005. They demonstrated a higher incidence of SLE when using SLICC criteria (58 cases) compared to ACR criteria (44 cases), mostly due to cases of isolated lupus nephritis, serologic abnormalities, and non-scarring alopecia.

Furst et al. (7) evaluated the incidence and prevalence of SLE in a large national managed-care claims database in the US using historical data. The incidence and prevalence of SLE was similar to previous estimates (table 1).

Lupus worldwide

Other than the recently published data from the Michigan Lupus Epidemiology and Surveillance Registry (8) which described a 2.1-fold higher incidence of SLE among Arab-Americans compared to non-Arab Caucasians and African Americans (8), little was known about the epidemiology of SLE among the Arab population worldwide or in the Middle East. Al Dhanhani et al. (9) studied the incidence and prevalence of SLE in the United Arab Emirates. The age-standardized incidence over the four-year period was 8.6 per 100,000/ year (table 2). The incidence rates described were similar to the ones reported in the Michigan Lupus Epidemiology and Surveillance Registry for the Arab population (7.6 per 100,000 person-years).

In Europe, Schneider et al. (10) used age-specific and sex-specific claims data to estimate the incidence of SLE in the German population (table 2). The estimated incidence rates of SLE were at the lower end of other estimates from comparable European countries, with the incidence rate for German women being less than half of the French rate (11).

Otsa et al. (12) estimated the incidence and point prevalence of SLE in Estonia by extracting SLE ICD-10 codes for individuals older than 20 years of age from the Estonian Health Insurance Fund database (table 2). The reported SLE incidence in Estonia was lower than in countries with similar prevalence, presumably due to the use of a lower age limit of 20 years as a study inclusion criterion.

In southern Sweden, Ingvarsson et al. (13) (table 2) reported a decrease in the incidence rate of SLE over a period of 26 years, particularly in middle-aged women, while disease phenotype remained unchanged. The prevalence of SLE increased slowly over the same period.

In Denmark, Hermanssen et al (14) (table 2) in an analysis of the Danish National registry reported an incidence rate for SLE of 2.35 per 100,000. Sex-specific incidence rates of SLE and of lupus nephritis peaked later in life among men than among women.

The first estimates of SLE incidence and prevalence on the island of Crete were reported (15) (table 2). While the incidence of SLE and lupus nephritis remained stable over the study period (1999–2013), prevalence increased.

A retrospective cohort study in the United Kingdom (16) (table 2) using the Clinical Practice Research Datalink showed a decline in the annual SLE incidence of 1.8% while in contrast the prevalence increased from 64.9 per 100,000 in 1999 to 97.04 in 2012. There was regional variation in both incidence and prevalence. People of Afro-Caribbean ethnicity had the highest incidence and prevalence.

In a systematic review of worldwide incidence and prevalence of SLE (17), the highest estimates of incidence and prevalence of SLE were in North America (23.2/100 000 person-years and 241/100 000, respectively). Lower incidences of SLE were reported in Africa and Ukraine (0.3/100 000 person-years), and the lowest prevalence was in Northern Australia (0 cases in a sample of 847 people). Women were more frequently affected than men for every age and ethnic group. People of African ethnicity had the highest incidence and prevalence of SLE, whereas those with Caucasian had the lowest incidence and prevalence. There appeared to be an increasing trend of SLE prevalence with time.

Mortality in SLE

A population-based study (18) using the National Vital Statistics System reported data on SLE deaths from 1968 through 2013. After an initial decrease between 1968 and 1975, SLE mortality increased annually for 24 years, followed by a sustained decrease for 14 years starting in 1999. On the other hand, all-cause mortality decreased throughout the study period. Residence in the West conferred the highest SLE mortality risk in all racial/ethnic groups except Caucasians, who had the highest risk in the South. Residence in the Northeast conferred the lowest mortality risk regardless of sex or ethnicity.

Costenbader et al. (19) analyzed Medicaid claims data and reported higher SLE mortality rates per 1000 patient-years among Native American (27.52), Caucasian (20.17), and African American (24.13) patients and were lower among Hispanic (7.12) or Asian (5.18) patients.

In a general population based study in the United Kingdom, SLE patients were shown to have nearly double the premature mortality risk of their peers (20).

Lee et al. (21) performed a meta-analysis of studies examining all-cause and cause-specific standard mortality rates (SMR) in SLE. All-cause standard mortality rates were increased 2.6 fold in SLE patients. The risk of mortality was significantly increased for mortality due to renal disease (SMR 4.689), cardiovascular disease (SMR 2.253), and infection (SMR 4.980), but not due to cancer (SMR 1.163).

Impact of family history

A study from Denmark (22) identified hospitalized patients with SLE over a period of 36 years and coupled them with their relatives through the Civil Registration System followed

by identification of twins using the Danish twin registry. Hazard ratios of SLE were high among first-degree (HR = 10.3) and second- or third-degree relatives of SLE patients (HR = 3.60). Risk of other autoimmune diseases was significantly increased both among SLE-affected first-degree (HR = 2.08) and second- or third-degree relatives (HR = 1.38).

In a separate publication (23), same authors reported a lower SLE twin concordance in Denmark than previously reported. Among seven monozygotic, eight same-sex dizygotic and five opposite-sex dizygotic twin pairs, one monozygotic and one dizygotic same-sex pair were concordant for SLE. This corresponded to proband and pairwise concordance rates of 25.0% and 14.3% for monozygotic twins, and proband and pairwise concordance rates of 7.7% for dizygotic twins.

Impact of age and gender

The RELESSER cohort data on gender differences among SLE patients (24) showed men to be diagnosed at a more advanced age than women. Men also had more cardiovascular comorbidities and were hospitalized more frequently. Men were more likely to lose weight, have lupus nephritis, lymphadenopathy, splenomegaly, and pulmonary fibrosis. Female patients were more likely to have inflammatory rash, alopecia, Raynaud's phenomenon, and arthritis.

A prospective single-center cohort in South Korea followed 133 children and 979 adults with SLE over a period of 14 years (25). Children with SLE had a higher number of cumulative ACR criteria and a higher adjusted mean SLE Disease Activity Index, but there was no difference in SLICC/ACR damage index. Immunosuppressants were used more frequently by children with SLE. The standardized mortality rate in pediatric SLE was 18.8, compared to 2.9 in adult SLE, a highly statistically significant difference.

Malignancy risk in SLE

Several studies examined the malignancy risk among SLE patients. A retrospective nested case-control study (26) which included 14842 patients in Taiwan analyzed the risk of malignancy among SLE patients on azathioprine, cyclophosphamide, methotrexate, hydroxychloroquine and glucocorticoids. A total of 330 patients developed a malignancy. The top five types of cancers were breast (16.9%), hematological (11.7%), colorectal (11.0%), lung (10.6%) and hepatobiliary (10.4%) cancers. The adjusted analyses showed an association of a higher cumulative cyclophosphamide dose (OR = 1.09) and lower hydroxychloroquine dose (OR = 0.93, 95% CI: 0.90, 0.97) with cancer risk in comparison with the controls.

A retrospective case-control study (27) included 40011 patients with an ICD-9 coded diagnosis of primary autoimmune disease, 311 of which had a concomitant coded diagnosis of myelodysplastic syndrome or acute myeloid leukemia. Among the 86 patients who met inclusion criteria, 12 had systemic lupus erythematosus. Patients on azathioprine had an odds ratio of 7.05 for development of a myeloproliferative syndrome ($P < .001$). Methotrexate (OR, 0.60), mercaptopurine (OR, 0.62), and mycophenolate mofetil (OR,

0.66) had favorable ORs that were not statistically significant. No association was found for anti-tumor necrosis factor agents.

Wadstrome et al. (28) examined the risk of cervical neoplasia in women with SLE in Sweden. There was an increased risk of cervical neoplasia in women with SLE compared with the general population (HR= 2.12). The subcohort treated with immunosuppressants was at highest risk of cervical neoplasia, compared with those treated with antimalarials.

Cardiovascular disease in SLE

Hermassen et al. (29) in a nationwide, population-based cohort study demonstrated a significantly higher risk of myocardial infarction and cardiovascular mortality in SLE patients with lupus nephritis compared to SLE patients without lupus nephritis ((HR=18.3 vs HR=2.2 for myocardial infarction and HR=7.8 vs HR=1.6 for cardiovascular mortality). The higher risk of stroke in SLE was not significantly affected by the presence of lupus nephritis.

Using data from the Swedish National Patient Register, Arkema et al. (30) studied the occurrence of ischemic and hemorrhagic stroke in patients with SLE. The relative risk of ischemic stroke in SLE was more than doubled compared with the general population (HR= 2.2), and importantly, the highest relative risks were observed within the first year after SLE diagnosis (HR=3.7).

To study ethnic differences in the cardiovascular risk among patients with SLE, Barbhaiya et al. (31) analyzed Medicaid data from 2000 to 2010 and identified 65788 SLE patients- 93.1% were women and 42% were black, 38% were white, 16% were Hispanic, 3% were Asian, and 1% were American Indian/Alaska Native. The risk of cardiovascular events was increased among blacks (HR 1.14) compared to whites, while Hispanics and Asians had a lower risk of MI (HR 0.61 and HR 0.57, respectively). Blacks and Hispanics had a higher risk of stroke (HR 1.31 and HR 1.22, respectively).

CONCLUSION

Over the past year we have gained more insight into the worldwide incidence and prevalence of SLE. Data from the California Lupus Surveillance Project and the Manhattan Lupus Surveillance Program allowed estimation of incidence and prevalence among Hispanics and Asians who had a higher incidence and prevalence of SLE compared to Caucasians, but lower than African Americans. In Germany, the SLE incidence rate for German women was less than half of the French rate. A decline in the annual SLE incidence of 1.8% was observed in the United Kingdom, while in contrast the prevalence increased by 50% over a period of 15 years ending with 2012.

Despite advances in treatment, standardized mortality rates in SLE remain three times higher than in the general population. The risk of mortality is significantly increased for mortality due to renal disease, cardiovascular disease, and infection.

SLE is a risk factor for cervical neoplasia, in particular for premalignant cervical lesions. The risk is highest among patients on immunosuppressants. Treatment-wise, azathioprine

was shown to increase the risk of acute myeloid leukemia or myelodysplastic syndrome 7-fold in an autoimmune population, but the study included only a small number of SLE patients.

Lupus nephritis was shown to be an important cardiovascular risk factor with a hazard ratio nine times higher compared to SLE patients without lupus nephritis. SLE doubles the risk of stroke with the highest relative risk for of stroke observed within the first year after diagnosis.

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Table 1

Lupus incidence and prevalence in the USA- a 2017 update

Authors	Region	Study type	Number of SLE cases	Ethnic distribution	Age	Gender	Study period	Incidence overall (cases per 100000 persons-years)	Incidence by ethnicity (cases per 100000 persons-years)	Prevalence (cases per 100000)	Prevalence by ethnicity (cases per 100000)
Somers et al. (1)	Southeastern Michigan	Population survey	2139	African American(n=1219) Caucasian(820) Hispanic(39)	All	8.5% males	2002–2004	5.5 (5.0–6.1)	African American 7.9 (6.9–9.1) Caucasian 3.7 (3.1–4.3)	72.8 (70.8–74.8)	African American 111.6 (107.7–115.6) Caucasian 47.5 (45.5–49.7)
Lim et al. (2)	Georgia	Population survey	1,446	African American (1,094) Caucasian (328)	All	13.5% males	2002–2004	6.9 (6.2,7.7)	African American 10.7 (9.5,12.1) Caucasian 3.3 (2.7,4.2)	92.1 (87.4,97)	African American 147.5 (139.2,156.4) Caucasian 43.1 (38.5,48.1)
Dall'Era et al. (4)	San Francisco county	Population survey	1,257	Caucasian (n = 294), Asian/Pacific Islander (n = 290), African American (n = 160), Hispanic (n=118), American Indian/Alaskan native (n = 4);	All	10.5% male	2007–2009	5.2 (4.3–6.2)	African American 16.0 (11.1–23.3) Caucasian 3.3 (2.4–4.4) Asian/ Pacific Islander 4.6 (3.3–6.3) Hispanic 5.6 (3.6–8.7)	96.8 (90.2–103.9)	African American 261.0 (222.3–306.5) Caucasian 64.9 (57.8–72.8) Asian/Pacific Islander 102.5 (91.3–115.1) Hispanic 110.5 (93.0–131.3)
Izmirly et al. (5)	New York county	Population survey	1,078	307 non-Hispanic white, 282 non-Hispanic black, 344 Hispanic, 111 non-Hispanic Asian	All	9.3% male	2007–2009	6.0 (4.6–7.4)	African American 10.1 (9.1–11.0) Caucasian 5.6 (4.2–7.1) Asian 5.4 (3.3–7.5)	75.9 (70.6–81.2)	African American 133.1 (130.6–135.7) Caucasian 51.4 (45.0–57.7)

Authors	Region	Study type	Number of SLE cases	Ethnic distribution	Age	Gender	Study period	Incidence overall (cases per 100000 persons-years)	Incidence by ethnicity (cases per 100000 persons-years)	Prevalence (cases per 100000)	Prevalence by ethnicity (cases per 100000)
									Hispanic 4.1 (3.8–4.5)		Asian 75.5 (66.0–85.0) Hispanic 84.6 (83.8–85.3)
Ferucci et al.(3)	Indian Health Service	Population survey	285	American Indian/Alaska Native only	All	11.9% male	2007–2009	7.4 (5.1–10.4)	N/A	178 (157–200)	N/A
Ungprasert et al(6)	Olmstead County, MN	Hospital and clinical records	58 (SLICC) 44 (ACR)	African American 7% Caucasian 86%	Over 18	16% (SLICC) 7%(ACR)	1993–2005	4.9 (SLICC) 3.7 (ACR97)	N/A	N/A	N/A
Furst et al. (7)	USA	Claims data	15396	N/A	Over 18	16% male	2003–2008	7.22	N/A	102.94	N/A

Table 2

Lupus incidence and prevalence worldwide- a 2017 update.

Authors	Region	Study type	Number of SLE cases	Ethnicity	Age	Gender distribution	Study period	Incidence (cases per 100000 persons-years)	Incidence by ethnicity (cases per 100000 persons-years)	Prevalence (cases per 100000)	Prevalence by ethnicity (cases per 100000)
Al Dhanhani et al. (9)	Al Ain region, United Arab Emirates	Hospital records, laboratory results, histopathology	15	Arab	All	20% male	2009–2012	8.6 (4.2–15.9)	N/A	103 (84.5– 124.4)	N/A
Brinks et al. (10)	Germany	Population survey	845	N/A	All	19.5% male	2002	Women: 3.6 (2.9 to 4.3) men: 2.2 (1.0 to 3.4)	N/A	N/A	N/A
Otsa et al. (12)	Estonia	Population survey	677	N/A	Adults over 20 years of age	10.5% male	2006–2010	1.4–1.7	N/A	37–40	N/A
Ingvarsson et al. (13)	Southern Sweden	Diagnosis registries, hospital records, laboratory databases	174	N/A	All	14.9% male	1981–2006	3.9 (2.1–5.5)	N/A	55–65	N/A
Hermanssen et al. (14)	Denmark	Population survey	1644	N/A	Adults over 18 years of age	14.3% male	1995–2011	2.35 (2.24–2.49)	N/A	45.2 (43.3–47.4)	N/A
Gergianaki et al. (15)	Crete	Chart review	750	N/A	Older than 15 years of age	7% male	1990 – 2011	8.6 (8.0 to 9.0)	N/A	123.4 (113.9 −132.9	N/A
Rees et al. (16)	United Kingdom	Retrospective cohort	7732	Caucasian 1700 African/ Carribean 128 Indian 45	All	14% male	1999–2012	4.91 (4.73 −5.09)	Caucasian 6.73 (6.35 to 7.14) African/ Carribean 31.46 (22.48 to 44.03) Indian 9.90 (6.32 to 15.53)	97.04 (94.19 to 99.94	Caucasian 134.53 (128.21 to 141.08) African/ Carribean 517.51 (398.54 to 660.84) Indian 193.09

Authors	Region	Study type	Number of SLE cases	Ethnicity	Age	Gender distribution	Study period	Incidence (cases per 100000 persons-years)	Incidence by ethnicity (cases per 100000 persons-years)	Prevalence (cases per 100000)	Prevalence by ethnicity (cases per 100000)
											(140.84 to 258.37)