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The worldwide incidence and prevalence of systemic lupus erythematosus: a systematic review of epidemiological studies

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Abstract

Objectives. The aim was to review the worldwide incidence and prevalence of SLE and variation with age, sex, ethnicity and time.

Methods. A systematic search of MEDLINE and EMBASE search engines was carried out using Medical Subject Headings and keyword search terms for Systemic Lupus Erythematosus combined with incidence, prevalence and epidemiology in August 2013 and updated in September 2016. Author, journal, year of publication, country, region, case-finding method, study period, number of incident or prevalent cases, incidence (per 100 000 person-years) or prevalence (per 100 000 persons) and age, sex or ethnic group-specific incidence or prevalence were collected.

Results. The highest estimates of incidence and prevalence of SLE were in North America [23.2/100 000 person-years (95% CI: 23.4, 24.0) and 241/100 000 people (95% CI: 130, 352), respectively]. The lowest 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. Incidence peaked in middle adulthood and occurred later for men. People of Black ethnicity had the highest incidence and prevalence of SLE, whereas those with White ethnicity had the lowest incidence and prevalence. There appeared to be an increasing trend of SLE prevalence with time.

Conclusion. There are worldwide differences in the incidence and prevalence of SLE that vary with sex, age, ethnicity and time. Further study of genetic and environmental risk factors may explain the reasons for these differences. More epidemiological studies in Africa are warranted.

Key words: incidence, prevalence, epidemiology, systemic lupus erythematosus, systematic review

CLINICAL SCIENCE

Rheumatology key messages

- There is wide geographical variation in the reported incidence and prevalence of SLE.
- Males with SLE have an older peak age of incidence and prevalence compared with females.
- There appears to be a trend of increasing prevalence of SLE with time.

Introduction

Systemic Lupus Erythematosus (SLE) is a chronic autoimmune disease with a varying clinical phenotype. It is known to affect women more frequently than men, with a ratio of approximately six women to every one man [1]. The aetiology of SLE is not fully understood, but both genetic predisposition and environmental triggers are believed to be involved [2]. Studying the epidemiology of SLE allows us to identify and explore changes in potential

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risk factors for the disease and allows planning of health services in response to overall disease burden [3]. A review of the incidence and prevalence of SLE was last published in 2006 by Danchenko *et al.* [4] and found marked disparities in incidence and prevalence worldwide. This was attributed to both true geographical variation and variation in study design. It could be a result of differences in the age and ethnic mix between populations, the definition of SLE used or, as found in some studies in the same population, a change in the incidence and prevalence of SLE with time [1, 5–7]. The aim of this study was to review the current literature published on the incidence and prevalence of SLE throughout the world.

Methods

A systematic literature review was undertaken. The search strategy used both Medical Subject Headings (MeSH) and keyword search terms for Systemic Lupus Erythematosus combined with MeSH and keyword terms for incidence and epidemiology, followed by prevalence and epidemiology (see supplementary Table S1, available at *Rheumatology* Online, for search strategy). The databases searched were Ovid MEDLINE from 1946 to August 2013 and EMBASE from 1974 to August 2013. All articles were downloaded into Endnote software and were selected on the basis of title and then abstract for full review. Hand-searching of citations also occurred. Articles were included if they were written in English or French language and were regarding humans. Exclusion criteria were review articles, conference proceedings, abstracts or editorials, articles in press, articles involving drug-induced lupus or neonatal lupus, and those solely regarding paediatric patients or a subtype of SLE, such as LN or discoid lupus. Searches were updated in September 2016. Table 1 shows the number of articles retrieved from each database in August 2013 and the additional articles added in September 2016.

Information on author, journal, year of publication, country, region, case-finding method, study period, number of incident or prevalent cases, incidence (per 100 000 person-years) or prevalence (per 100 000 persons) was collected by F.R. In addition, any age, sex or ethnic group-specific incidence or prevalence rates reported were collected. Age-adjusted or standardized results were presented whenever available. PRISMA guidelines were used.

Results

Incidence

Geography

Table 2 and Fig. 1A summarize the reported worldwide incidence estimates of SLE. Figure 1A uses the most recent estimates from Table 2. There was worldwide variation, with the highest incidence reported in North America (23.2/100 000 person-years, 95% CI: 22.4, 24.0) [8] and the lowest incidences reported in Africa (0.3/100 000 person-years) [9] and Ukraine (0.3/100 000 person-years, 95% CI: 0.0, 1.5) [10]. In general, European countries had a lower incidence of SLE, whereas Asia, Australasia and the Americas had a higher incidence. The most frequent methods for case-finding were local secondary care hospital-based outpatient lists or discharge registries, or National Health Insurance databases.

Age and sex

In all studies reviewed, females had a higher incidence of SLE compared with males. The sex ratio ranged from 2:1 [36] to 15:1 [46]. As an example, Somers *et al.* [31] estimated the UK incidence to be 7.89/100 000 person-years (95% CI: 7.46, 8.31) for females compared with 1.53/100 000 person-years (95% CI: 1.34, 1.71) for males. This higher incidence in females remained true for every age group, although the ratios were smaller at both extremes of age.

In the majority of studies, there was a peak age of incidence before declining. In females, the peak age ranged from the third to seventh decades of life. For males, the peak incidence was usually later, in the fifth to seventh decades. Three selected studies taken from three different geographical regions demonstrate this in Fig. 2A.

Ethnicity

In studies that reported differences between ethnic groups [1, 8, 21, 29, 33, 35, 37, 41, 42, 58, 59], incidence rates

Table 1 Summary of literature search

Search term	Database	Number of articles retrieved	Number of articles after removing duplicates	Number of articles selected for review on the basis of title and abstract	Number of articles selected for inclusion after reading the full text article, including additional articles found by hand searching	Number of additional articles selected on updated search in September 2016
Incidence	Medline Embase	542 1175	1617	76	46	11
Prevalence	Medline Embase	929 2290	2744	92	76	14

Table 2 Worldwide incidence of SLE

Continent	Country	References	Region	Case-finding method	Number of incident cases	Incidence per 100 000 person-years (95% CI) [study year]
Europe	Denmark	Voss *et al.* [5]	Funen	Hospital and community records	127	1.0 (0.3, 2.9)[a] [1980] 3.6 (2.0, 6.1)[a] [1994]
		Laustrup *et al.* [11]	Funen	Hospital and community records	35	1.0 (0.3, 2.7)
		Hermansen *et al.* [12]	National	National patient registry	1644	2.35 (2.24, 2.49)
	France	Arnaud *et al.* [13]	National	National health insurance database	1931	3.32
	Finland	Elfving *et al.* [14]	Northern Savo	Hospital and community records	7	3.6 (3.0, 4.2)[a]
	Greece	Alamanos *et al.* [15]	North-west	Hospital records	178	1.9 (1.5, 2.3)[a]
	Iceland	Gudmundsson *et al.* [16]	National	Hospital registers	76	3.3
	Italy	Govoni *et al.* [17]	Ferrara	Hospital records	2000: 7	2.0
					2001: 4	1.2
					2002: 9	2.6
		Tsioni *et al.* [18]	Valtrompia	Hospital and community records	9	2.0 (0.9, 3.8)
	Norway	Nossent [19]	North	Hospital records	83	2.9 (2.4, 3.3)[a]
		Eilertsen *et al.* [20]	North	Hospital records	58	3.0 (2.0, 4.0)
		Lerang *et al.* [21]	Oslo	Hospital records	116	3.0 (2.4, 3.5)
	Spain	López *et al.* [22]	Asturias	Hospital records	116	2.2 (1.8, 2.5)
		Gómez *et al.* [23]	Asturias	Hospital records	–	1.9 (1.1, 2.7)
		Alonso *et al.* [24]	Lugo	Hospital records	150	3.6 (3.0, 4.2)[a]
	Sweden	Leonhardt [7]	Malmö	Hospital records	16	1.0[a]
		Eyrich *et al.* [25]	Halmstad	Hospital records	41	1.8 [1957, 1964] 3.0 [1964, 1971]
		Jonsson *et al.* [26]	Lund and Orup	Hospital and community records	39	4.0 (1.6, 6.4)[a]
		Ståhl-Hallengren *et al.* [6]	Lund and Orup	Hospital and community records	41	4.8
		Ingvarsson *et al.* [27]	Lund and Orup	Hospital and community records	55	2.8 (1.4, 4.2)
	UK	Hopkinson *et al.* [28]	Nottingham	Hospital records	23	4.0 (2.3, 5.6)[a]
		Johnson *et al.* [29]	Birmingham	Hospital records	33	3.8 (2.5, 5.1)
		Nightingale *et al.* [30]	Whole UK	CPRD	390	3.0 (2.7, 3.3)
		Somers *et al.* [31]	Whole UK	CPRD	1638	4.7 (4.5, 4.9)[a]
		Rees *et al.* [1]	Whole UK	CPRD	2740	4.9 (4.7, 5.1)
North America	Canada	Bernatsky *et al.* [32]	Quebec	Physician billing database	219	3.0 (2.6, 3.4)
				Hospitalization database	203	2.8 (2.6, 3.0)
	USA	Siegel *et al.* [33]	New York and Alabama	Hospital records	New York: 98	1.9
					Alabama: 63	1.0
		Fessel [34]	San Francisco	Hospital records	74	7.6
		Hochberg [35]	Baltimore	Hospital records	302	4.6[a]
		Michet *et al.* [36]	Minnesota	Hospital records and death certificates	25	1.8 (1.1, 2.5)[a]
		McCarty *et al.* [37]	Pennsylvania	Community and hospital records	191	2.4 (2.1, 2.8)[a]

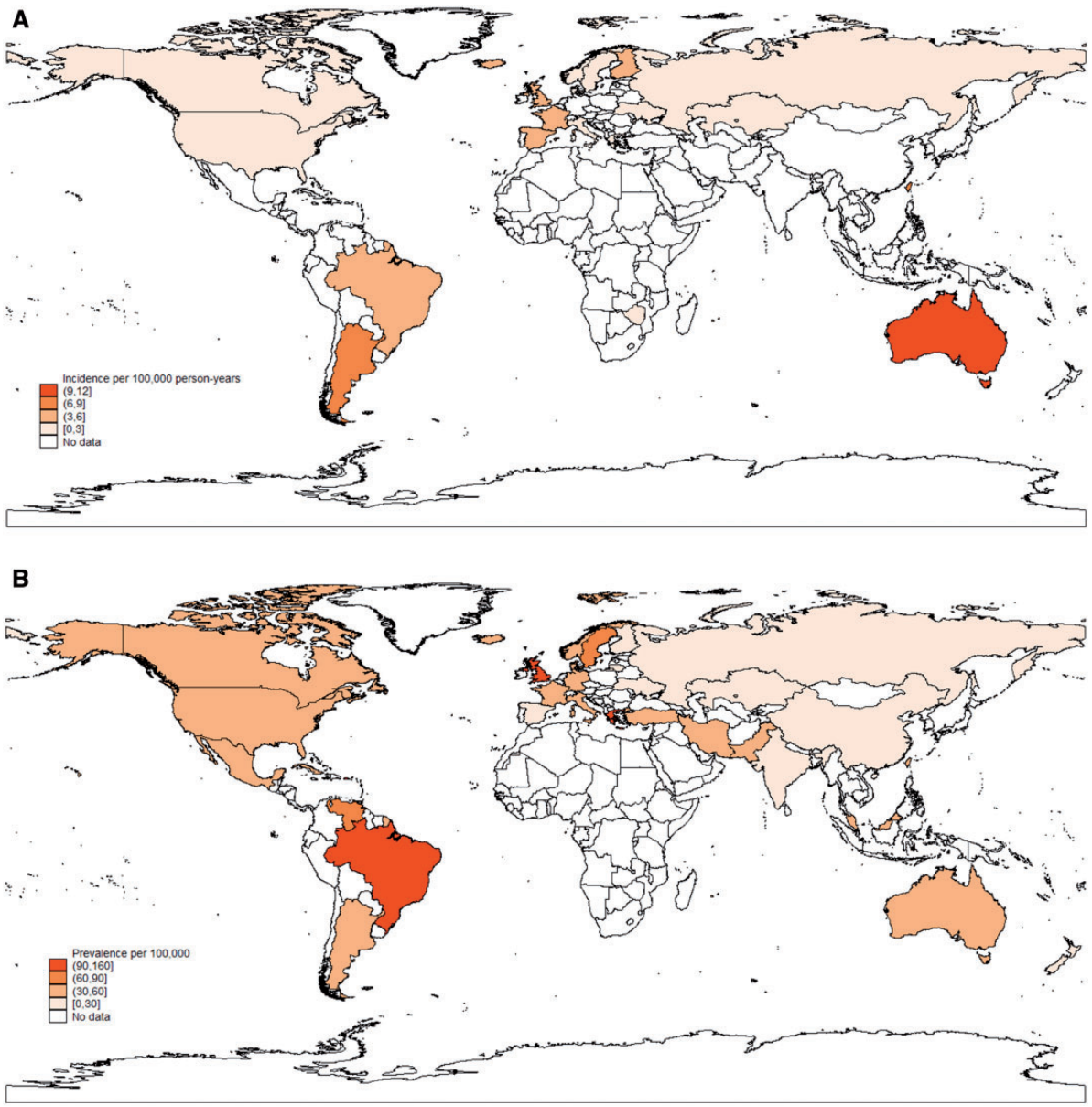
(continued)

Table 2 Continued

Continent	Country	References	Region	Case-finding method	Number of incident cases	Incidence per 100 000 person-years (95% CI) [study year]
		Uramoto *et al.* [38]	Minnesota	Hospital records	48	5.6 (3.9, 7.2)[a]
		Naleway *et al.* [39]	Wisconsin	Medical records	44	5.1 (3.6, 6.6)[a]
		Feldman *et al.* [8]	Whole US	Medicaid database	3490	23.2 (22.4, 24.0)
		Furst *et al.* [40]	Whole US	Medical claims database	1557	7.2 (6.8, 7.7)[a]
		Lim *et al.* [41]	Georgia	Georgia Lupus registry	267	5.6 (5.0, 6.3)[a]
		Somers *et al.* [42]	Michigan	Medical records	399	5.5 (5.0, 6.1)[a]
		Jarukitsopa *et al.* [43]	Minnesota	Rochester epidemiology project database	45	2.9 (2.0, 3.7)
Central America	Caribbean	Nossent [44]	Curaçao	Medical records	68	4.6 (0.4, 8.8)
		Deligny *et al.* [45]	Martinique	Medical records	180	4.7 (2.5, 6.9)
		Flower *et al.* [46]	Barbados	National hospital-based SLE registry	183	6.3 (5.4, 7.3)[a]
South America	Argentina	Scolnik [47]	Buenos Aires	Private medical care database	68	6.3 (4.9, 7.7)
	Brazil	Pereira Vilar *et al.* [48]	Natal city	Hospital records	43	8.7 (6.3, 11.7)
		Nakashima *et al.* [49]	Cascavel	Medical records	14	4.8
Africa	Zimbabwe	Taylor *et al.* [9]	Bulawayo and Harare	Hospital records	22	0.3
Asia	China	Mok *et al.* [50]	Hong Kong	University hospital database	–	3.1
	Kazakhstan	Nasonov *et al.* [10]	Semey	Hospital records	4	1.3 (0.4, 3.4)[a]
	Russia	Nasonov *et al.* [10]	Kursk and Yaroslavl	Hospital records	12	1.2 (0.6, 2.1)[a]
	Ukraine	Nasonov *et al.* [10]	Vinnitsa	Hospital records	1	0.3 (0.0, 1.5)[a]
	South Korea	Shim *et al.* [51]	National	National Health Insurance database	1398	2.8 (2.7–2.9)[a]
	Taiwan	Chiu *et al.* [52]	National	National Health Insurance database	12 789	8.1
		Kang *et al.* [53]	National	National Health Insurance database	758	3.3
		Yu *et al.* [54]	National	National Health Insurance database	671	8.4 (7.7, 9.0)
		Yeh *et al.* [55]	National	Catastrophic illness database	6675	4.9
		See *et al.* [56]	National	National Health Insurance database	358	7.2 (6.5, 8.0)
Australasia	Australia	Anstey *et al.* [57]	Northern Territory	Hospital records	13	11

[a]Age standardized. CPRD: UK Clinical Practice Research Datalink.

FIG. 1 The global incidence **(A)** and prevalence **(B)** of SLE (most recent estimates used)

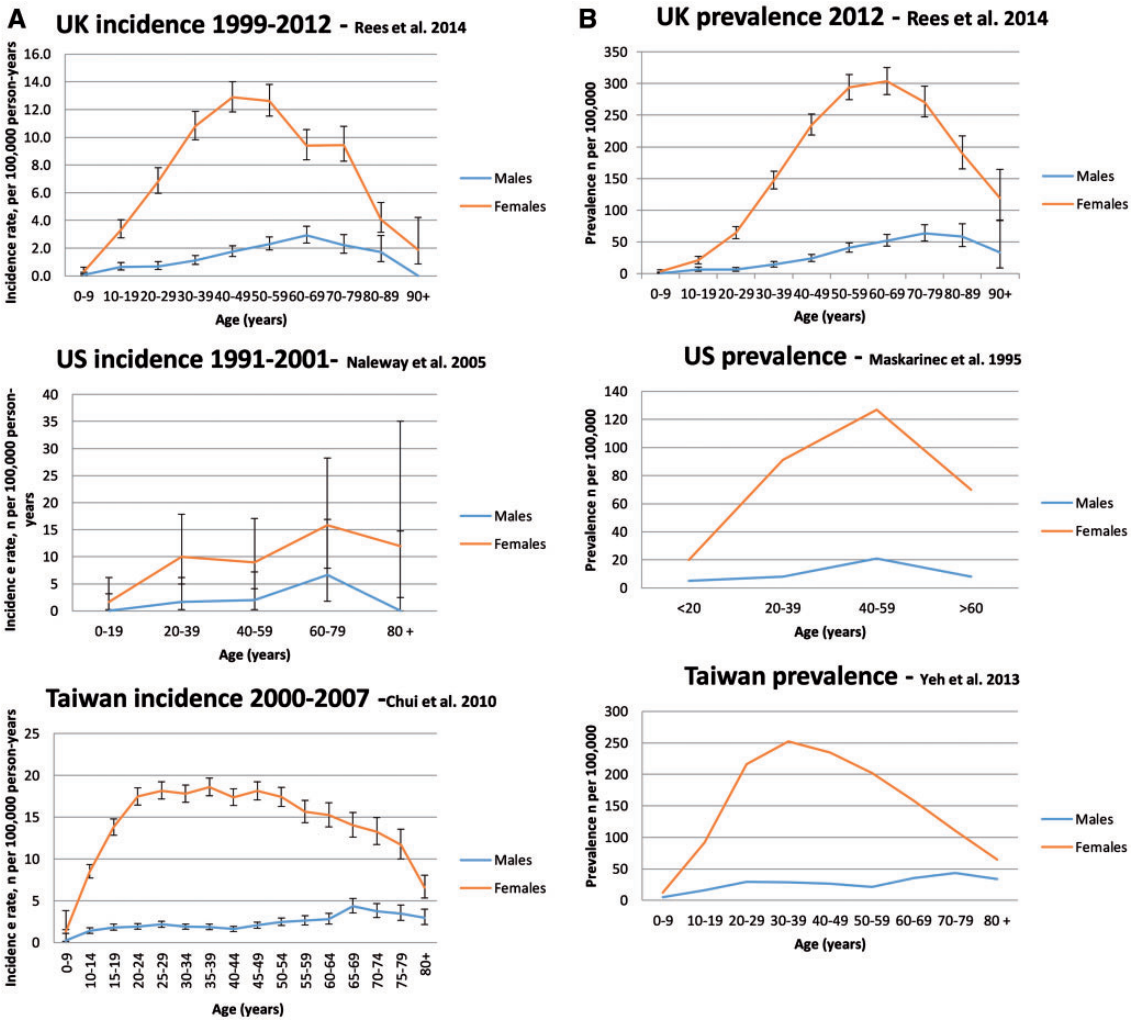


were highest in Black populations and lowest in Caucasians. Asian and Hispanic ethnic groups were intermediate. For example, in the UK, Hopkinson *et al.* [59] published race-specific incidence figures for Nottingham, with Afro-Caribbeans highest at 31.9/100 000 person-years, Asians 4.1/100 000 person-years and Whites 3.4/100 000 person-years. In North America, Native American Indians also had higher incidence rates than the White population. This was demonstrated in the study by Feldman *et al.* [8], where the incidence in native American Indians was 30.0/100 000 person-years (95% CI: 22.5, 39.9), which was similar to that of Black or African Americans [31.2/100 000 person-years (95% CI: 29.6, 32.9)] and significantly higher than for Whites [18.0/100 000 person-years (95% CI: 17.0, 19.0)] and Asians [16.7/100 000 person-years (95% CI: 13.9, 20.0)]. In the same study, the incidence in Hispanics was 22.2/100 000 person-years (95% CI: 20.4, 24.2). A study specifically focusing on native American Indians found that three tribes had a particularly high incidence of SLE, specifically the Crow, Arapahoe and Sioux tribes [60].

Temporal trend

There were a number of studies that examined the same population at risk over time, allowing us to examine the temporal trend (Fig. 3A). In the UK, Somers *et al.* [31]

FIG. 2 The incidence **(A)** and prevalence **(B)** of SLE stratified by age and sex in the UK, USA and Taiwan



showed a small but non-significant increase in the incidence in females over the 10-year period 1990–99, but not with males. However, Rees *et al.* [1] found a statistically significant decline in incidence from 1999 to 2012 of 1.8% per year. In the County of Funen in Denmark, Voss *et al.* [5] looked at the time periods 1980–84, 1985–89 and 1990–94. The respective incidence rates were 1.0 (95% CI: 0.6, 1.6), 1.1 (95% CI: 0.7, 1.7) and 2.5 (95% CI: 1.8, 3.3) per 100 000 person-years. Although not linear, there was a significant increase from the first to the last 5-year period. Although this could be a true increase, from 1 January 1993 an additional data source was available, thus increasing the number of cases identified. Alamanos *et al.* [15], in North-West Greece, showed an increasing trend from 1.41/100 000 person-years (95% CI: 0.99, 1.83) in 1982–86 to 2.19/100 000 person-years (95% CI: 1.78, 2.60) in 1997–2001, but this was not statistically significant. Finally, results from the Rochester Epidemiology project in Minnesota were published by Michet *et al.* [36] for the period 1950–79, when the incidence was 1.8/100 000 person-years (95% CI: 1.1, 2.5), followed by Uramoto *et al.* [38], who published data for 1980–92, when the incidence rate was 5.6/100 000 person-years (95% CI: 3.9, 7.2), and finally, Jarukitsopa *et al.* [43], who examined 1993–2005 and found the incidence rate had declined to 2.9/100 000 person-years (95% CI: 2.0, 3.7).

Prevalence

Geography

The prevalence of SLE by country is summarized in Table 3 and Fig. 1B. Figure 1B uses the most recent estimates from Table 3. The lowest prevalence was reported in a community study of 847 people in Yarrabah, North Queensland, Australia [61], where no cases were found. The highest prevalence was in a national survey in the USA [62], which reported a prevalence of 241/100 000

Table 3 Worldwide prevalence of SLE

Continent	Country	References	Study period	Region	Case-finding method	Prevalent cases	Prevalence, per 100 000 (95% CI) [year of study]
Europe	Denmark	Voss *et al.* [5]	1 January 1995	Funen	Hospital and community records	84	22.2[a]
		Laustrup *et al.* [11]	1 January 2003	Funen	Hospital and community records	109	28.3 (23.3, 34.2)
		Eaton *et al.* [63]	31 October 2006	National	National hospital patient registry	–	48
		Hermansen *et al.* [12]	31 Decmeber 2011	National	National hospital patient registry	1887	45.2 (43.3, 47.4)
	Finland	Helve [64]	December 1978	National	National hospital discharge database	1427	28
	France	Arnaud *et al.* [13]	2010	National	National Health Insurance database	27 369	40.8[a]
	Germany	Brinks *et al.* [65]	2002	National	National Health Insurance database	845	36.7 (34.3, 39.3)
	Greece	Alamanos *et al.* [15]	31 December 2001	North-West	Hospital records	193	38.1 (36.3, 39.9)[a]
		Anagnostopoulos *et al.* [66]	2008	Central	Postal survey	2	110 (110, 370)
	Iceland	Gudmundsson *et al.* [16]	1975–84	National	Hospital registers	86	35.9[a]
	Italy	Benucci *et al.* [67]	June 2002	Florence	Community survey	23	71 (49, 92)[a]
		Govoni *et al.* [17]	2002	Ferrara	Hospital records	201	57.9
		Sardu *et al.* [68]	July 2009	Southern Sardinia	Community records	–	81 (50, 124)
		Tsioni *et al.* [18]	31 December 2012	Valtrompia	Hospital and community records	44	39.2 (28.5, 52.6)
	Lithuania	Dadoniene *et al.* [69]	2004	Vilnius	Hospital records and community survey	76	16.2 (12.7, 20.3)
	Norway	Nossent [19]	1996	North	Hospital records	89	49.7 (44.3, 55)[a]
		Eilertsen *et al.* [20]	2007	North	Hospital records	114	64.1
		Lerang *et al.* [21]	1 January 2008	Oslo	Hospital records	238	52.8 (45.2, 58.4)
	Spain	López *et al.* [22]	31 December 2002	Asturias	Hospital records	367	34.1 (30.6, 37.6)
		Gómez *et al.* [23]	December 2003	Asturias	Hospital records	–	31.7 (28.3, 35.0)
		Alonso *et al.* [24]	31 December 2006	Lugo	Hospital records	150	17.5 (12.6, 24.1)[a]
	Sweden	Leonhardt [7]	1955 1958 1961	Malmö	Hospital records	–	2.9 4.5 6.0
		Nived *et al.* [70]	31 December 1982	Lund and Orup	Hospital and community records	61	39 (30, 48)

(continued)

Table 3 Continued

Continent	Country	References	Study period	Region	Case-finding method	Prevalent cases	Prevalence, per 100 000 (95% CI) [year of study]
		Ståhl-Hallengren *et al.* [6]	31 December 1986	Lund and Orup	Hospital and community records	121	42
			31 December 1991			162	68
		Simard *et al.* [71]	1 January 2010	National	National patient register	7929	(46, 85)
		Ingvarsson *et al.* [27]	31 December 2006	Lund and Orup	Hospital and community records	174	65
	Turkey	Çakır *et al.* [72]	–	Havsa	Community survey	10	57 (46, 70)[a]
	UK	Hochberg [73]	1981–82	Whole UK	Community medical record survey	20	6.5
		Samanta *et al.* [74]	1986–89	Leicester	Hospital records	50	26.1
		Hopkinson *et al.* [28]	30 April 1990	Nottingham	Hospital records	147	24.6 (20.6, 28.7)[a]
		Johnson *et al.* [29]	1992	Birmingham	Hospital records	242	27.7 (24.2, 31.2)
		Gourley *et al.* [75]	1 August 1993	Northern Ireland	Hospital records	408	25.4 (22.1, 28.7)[a]
		Nightingale *et al.* [76]	1992–98	Whole UK	CPRD	1538	25.0 (23.4, 26.7) [1992]
							40.7 (37.6, 43.8) [1998]
		Rees *et al.* [1]	1999–2012	Whole UK	CPRD	1875	65.0 (62.1, 67.9) [1999][a]
						4413	97.0 (94.2, 99.9) [2012][a]
North America	Canada	Peschken *et al.* [77]	1996	Manitoba	Medical records	257	22.1 (13.2, 32.4)
		Bernatsky *et al.* [32]	2003	Quebec	Physician billing and hospitalization databases	3825	44.7 (37.4, 54.7)[a]
	USA	Siegel *et al.* [58]	1959	New York	Hospital records	–	5
		Fessel [34]	1973	San Francisco	Hospital records	64	50.8
		Serdula *et al.* [78]	1975	Oahu, Hawaii	Hospital records	81	15.3[a]
		Michet *et al.* [36]	1 January 1980	Minnesota	Hospital records	20	40.0 (23.5, 57.5)
		Uramoto *et al.* [38]	1 January 1993	Minnesota	Hospital records	–	122 (97, 217)[a]
		Maskarinec *et al.* [79]	1989	Hawaii		454	41.8
		Post *et al.* [80]	1996	California	Postal survey	20	68.2
		Balluz *et al.* [81]	1997	Arizona	Hospital and community records	20	103 (56, 149)
		Ward [62]	1988–94	National	US National health survey	40	241 (130, 352)
		Naleway *et al.* [39]	2001	Wisconsin	Medical records	64	78.5 (59.0, 98.0)[a]
		Chakravarty *et al.* [82]	2000	California and Pennsylvania	Hospitalization databases	–	California: 107.6 (106.1, 109.2)[a]

(continued)

Table 3 Continued

Continent	Country	References	Study period	Region	Case-finding method	Prevalent cases	Prevalence, per 100 000 (95% CI) [year of study]
							Pennsylvania: 149.5 (146.9, 152.2)[a]
		Feldman *et al.* [8]	2000–04	National	Medicaid database	34339	143.7 (142.2, 145.3)
		Furst *et al.* [40]	2003–08	National	Medical claims database	15396	81.1 (78.5, 83.6) [2003] 102.9 (100.4, 105.5) [2008]
		Lim *et al.* [41]	2002	Georgia	Georgia Lupus registry	1156	73.0 (68.9, 77.4)[a]
		Somers *et al.* [42]	2002–04	Michigan	Medical records	2139	72.8 (70.8, 74.8)[a]
		Jarukitsopa *et al.* [43]	1 January 2006	Rochester, MN	Rochester epidemiology project database	72	53.5 (41.1, 65.9)
Central America	Caribbean	Nossent [44]	1 January 1990	Curaçao	Medical records	69	47.6 (34.1, 51.1)
		Deligny *et al.* [45]	1999	Martinique	Medical records	245	64.2 (56.2, 72.2)
		Molina *et al.* [83]	2003	Puerto Rico	Private health insurance database	877	159
		Reyes-Llerena *et al.* [84]	–	Havana, Cuba	WHO-ILAR COPCORD study	2	60 (10, 200)
		Flower *et al.* [46]	31 October 2009	Barbados	National hospital-based SLE registry	226	84.1 (73.5, 95.8)
	Mexico	Peláez-Ballestas *et al.* [85]	–	Five regions in Mexico	WHO-ILAR COPCORD study	–	60 (30, 100)[a]
South America	Argentina	Scolnik *et al.* [47]	1 January 2009	Buenos Aires	Private medical care database	75	58.6 (46.1, 73.5)
	Brazil	Rodrigues Senna *et al.* [86]	–	Montes Claros City	WHO-ILAR COPCORD study	3	98 (20, 280)
	Venezuela	Granados *et al.* [87]	2011	Monagos	WHO-ILAR COPCORD study	3	70 (10, 200)
Asia	China	Wigley *et al.* [88]	–	North (near Beijing) South (near Shantou)	WHO-ILAR COPCORD study	North: 3 South: 1	10 20
		Li *et al.* [89]	–	Beijing	Community survey	3	30 (0, 60)
	India	Malaviya *et al.* [90]	–	Delhi	Community survey	3	3.2 (0, 6.86)
	Iran	Davatchi *et al.* [91]	September 2005	Tehran city	WHO-ILAR COPCORD study	3	40
		Davatchi *et al.* [92]	September 2006	Five villages in NW Iran	WHO-ILAR COPCORD study	1	60 (6, 670)

(continued)

Table 3 Continued

Continent	Country	References	Study period	Region	Case-finding method	Prevalent cases	Prevalence, per 100 000 (95% CI) [year of study]
	Kazakhstan	Nasonov *et al.* [10]	31 December 2010	Semey	Hospital records	52	17.3 (12.9, 22.6)[a]
	Malaysia	Wang *et al.* [93]	1974–90	Kuala Lumpur	Hospital records	539	43
	Pakistan	Farooqi *et al.* [94]	–	North	WHO-ILAR COPCORD study	1	50
	Russia	Nasonov *et al.* [10]	31 December 2010	Kursk and Yaroslavl	Hospital records	79	7.7 (6.1, 9.7)[a]
	South Korea	Ju *et al.* [95]	2004–06	National	National Health Insurance database	9000–11000	18.8, 21.7
		Shim *et al.* [51]	2006–10	National	National Health Insurance database	10080	20.6 (20.2, 21.0) [2006]
						13316	26.5 (26.0, 27.0) [2010]
	Taiwan	Chou *et al.* [96]	–	Cu-Tien	Community survey	1	33
		Chiu *et al.* [52]	2000–07	National	National Health Insurance database	15463	42.2 [2000] 67.4 [2007]
		Kang *et al.* [53]	31 December 2005	National	National Health Insurance database	15753	69.3
		Yu *et al.* [54]	2000	National	National Health Insurance database	356	37.0 (10.0, 41.0)
		Yeh *et al.* [55]	2003 2008	National	Catastrophic illness database	133488	97.5
		See *et al.* [56]	2005	National	National Health Insurance database	435	43.5 (39.4, 47.6)
	Ukraine	Nasonov *et al.* [10]	31 December 2010	Vinnitsa	Hospital records	45	12.2 (8.9, 16.4)[a]
Australasia	Australia	Anstey *et al.* [57]	1January 1991	Northern Territory.	Hospital records	22	52
		Grennan *et al.* [97]	1993	Queensland Sydney	Hospital records	Queensland: 20	89
						Sydney: 3	13
		Bossingham [98]	1 August 1996 to 31 August 1998	Far North Queensland	Hospital records	108	45.3
		Minaur *et al.* [61]	January 2002	Yarrabah, North Queensland	WHO-ILAR COPCORD study	0	0
	New Zealand	Meddings *et al.* [99]	–	Dunedin	Hospital records	16	14.7
		Hart *et al.* [100]	1980	Auckland	Hospital records	136	17.6[a]

[a]Age standardized. CPRD: clinical practice research datalink; WHO-ILAR COPCORD: World Health Organization–ILAR Community Orientated Program for the Control of Rheumatic Diseases.

people (95% CI: 130, 352). The most frequent methods for case-finding were local secondary care hospital-based outpatient or discharge registries, National Health Insurance databases or community surveys, such as the World Health Organization–ILAR Community Orientated Program for the Control of Rheumatic Diseases (WHO-ILAR COPCORD).

Age and sex

In all studies, prevalence was highest among females, with a female to male ratio ranging between 1.2:1 [86] and 15:1 [46]. As an example, in Birmingham in the UK, Johnson *et al*. [29] found estimates of 49.6/100 000 (95% CI: 43.2, 56.1) for women compared with 3.6/100 000 (95% CI: 2.0, 6.0) for men in a hospital-based study. A further study in Birmingham, UK in 1996 aimed to identify undiagnosed cases of SLE in the community via a postal questionnaire sent to a random sample of 3500 women aged 18–65 years. This suggested a much greater prevalence in women of 200/100 000 (95% CI: 80, 412) [101] compared with the hospital-based study.

Prevalence curves by age had a similar distribution to that of the incidence data, but with a later peak age. Figure 2B shows the age- and sex-specific prevalence from three papers from selected countries from around the world. Summarizing studies from the UK, the peak age of prevalence was between 45 and 69 years for females and between 40 and 89 years for males [1, 76]. Most worldwide studies confirmed the delayed peak age of incidence in males apart from two studies from Scandinavia, which found a lower peak age in men [21, 70].

Ethnicity

Similar to the incidence data, Black ethnic groups had the highest reported prevalence of SLE, White groups the lowest and Asian and Hispanic groups were intermediate for both males and females. As an example, the prevalence in different ethnic groups in the UK is summarized in Table 4.

In addition to the studies in Table 4, a study of women aged 15–64 years in South London estimated the prevalence of SLE to be 177/100 000 (95% CI: 135, 220) in Afro-Caribbean people and 110/100 000 (95% CI: 58, 163) in West African people compared with 35/100 000 (95% CI: 26, 43) in White European people [103]. Studies from the USA have also confirmed the difference between Black and White populations [8, 33], with intermediate figures for Hispanic, Asian and native North Americans. A study from Hawaii had the greatest ethnic diversity [78]. Here, Chinese and native Hawaiian groups were most prevalent (24.1 and 20.4/100 000, respectively) and Whites least prevalent (5.8/100 000; 95% CI not given). In the same study, White people had a significantly older mean age of disease prevalence of 38.1 years, compared with 29.7 years overall.

Temporal trend

There appeared to be a trend for increasing prevalence with time (Fig. 3B). In the UK, the crude annual prevalence of SLE reported by Nightingale *et al.* [76] increased from 25/100 000 (95% CI: 23.4, 26.7) in 1992 to 40.7/100 000 (95% CI: 37.6, 43.8) in 1998. A subsequent study by Rees *et al.* [1] confirmed this trend and found that prevalence rose annually by 3.1% from 1999 to 2012, which was statistically significant. In Malmö, Sweden the prevalence rose from 2.9/100 000 in 1955 to 6.0/100 000 in 1961 [7] and in Lund and Orup from 39/100 000 (95% CI: 30, 48) on 31 December 1982 [70] to 68/100 000 on 31 December 1991 [6]. The same trend was found in Northern Norway [11, 20] and Minnesota [36, 38].

Discussion

There are five main findings from this systematic review: there is worldwide variation in the reported incidence and prevalence of SLE; in all nationalities, there is a female predominance; there is a peak age of incidence, which occurs in middle-aged adults; Black ethnic groups have the highest incidence and prevalence and White ethnic groups have the lowest; and there appears to be an increasing trend in the prevalence of SLE with time.

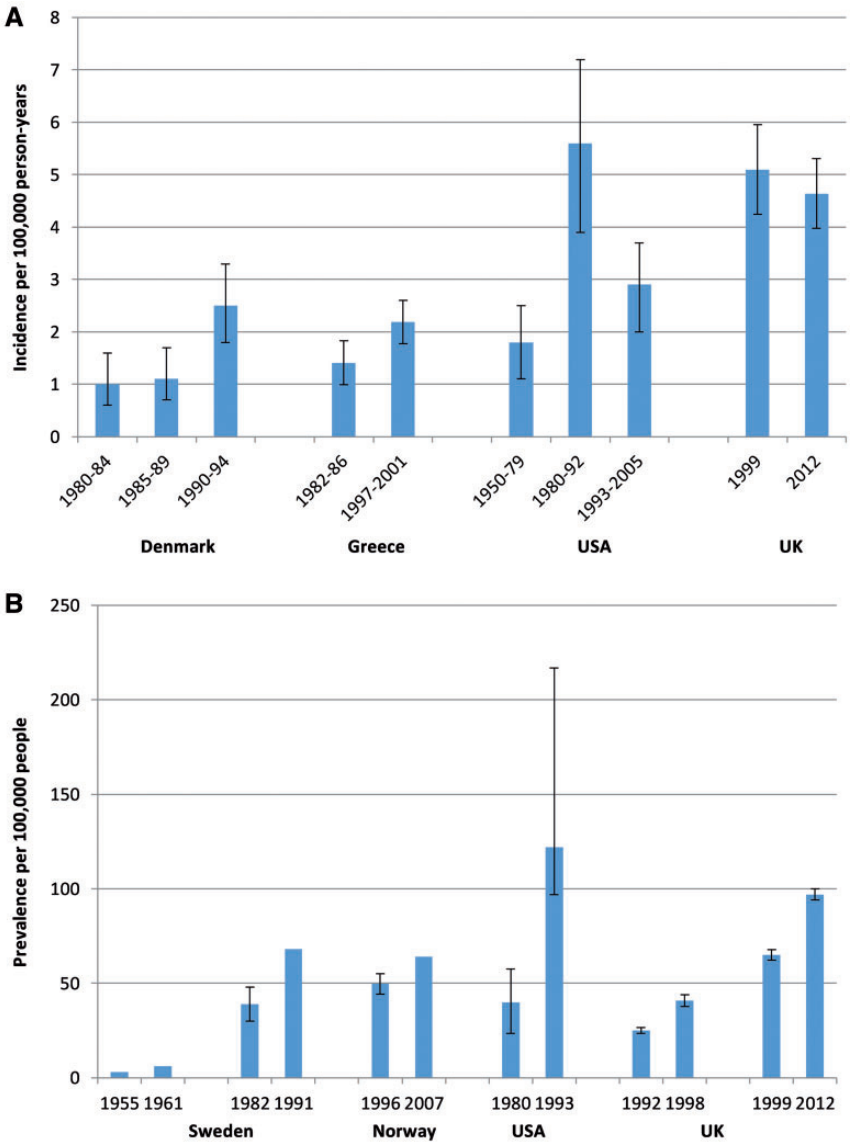
The geographical variation could reflect differences in the genetic mix of populations or variation in

Table 4 The prevalence of SLE in the UK by ethnicity

References	Region	Prevalence per 100 000 (95% CI)			
		Black	Asian	White	Chinese
Samanta *et al.* [102]	Leicester	–	40[a]	20[a]	–
Samanta *et al.* [74]	Leicester	–	64.0[a]	20.2	–
Hopkinson *et al.* [59]	Nottingham	207.0	48.8[a]	20.3[a]	92.9[a]
Johnson *et al.* [29]	Birmingham	197.2	96.5[a]	36.3[a]	–
Rees *et al.* [1]	National	African: 179.8 (125.2, 250.1) Caribbean: 517.5 (398.5, 660.8)	Indian: 193.1 (140.8, 258.4)	134.5 (128.2, 141.1)	188.39 (90.3, 346.5)

[a]Age-standardized.

Fig. 3 Temporal trend for the incidence **(A)** and prevalence **(B)** of SLE



environmental exposures; for example, countries nearer the equator are exposed to more ultraviolet radiation, which has been hypothesized to be an environmental trigger for SLE [104, 105]. The variation could also be attributable to differences in the epidemiological study methods used, the diagnosis rates of SLE in each country, the diagnostic criteria used, access to health care, access to immunology laboratory tests and differing thresholds for positive results, the decade the study was carried out, whether the rates were age adjusted and, if not age adjusted, the underlying population structures. For example, the incidence of SLE in Zimbabwe was one of the lowest worldwide. This may have been underestimated because the data were collected retrospectively, relied on the attendance of people with SLE at one of the study hospitals during the study period, it was not an age-adjusted rate, and life expectancy is lower in Zimbabwe such that the peak age of onset may exceed the average life expectancy. Likewise, the low prevalence found in Australia may be attributable to the fact that it was a small community survey of Australian Aboriginal people in Yarrabah, North Queensland and was underpowered to detect any SLE cases. The North American estimate of SLE incidence of 23.2/100 000 person-years may be overestimated because it is significantly higher than all the other USA estimates. This may be because it is an unadjusted rate or may reflect methodological differences rather than genetic or environmental differences in the population at risk. This study used the Medicaid database, which may have self-selected people with a

chronic disease such as SLE, who may be overrepresented in Medicaid, and hence increased the estimate. It should be emphasized that Fig. 1 used data from different decades and from studies using different case-ascertainment methods so should be interpreted with caution.

In common with other conditions that display autoimmune features, SLE is universally more common in females. This could relate both to possession of the double X chromosome and to differences in oestrogen levels, which modulate immune responses [106, 107]. Hormonal changes have been hypothesized to explain the peak incidence in women in young to middle adulthood compared with childhood and older adulthood. However, this explanation cannot fully explain why the peak in incidence extends into the post-menopausal age group [2] unless there is a longer latency between the rise in oestrogen levels, the triggering of the autoimmune pathway and the development of clinical disease in some women.

Incidence and prevalence peak in middle age. Most worldwide studies confirmed the delayed peak age of prevalence in males. Interestingly, two studies from Scandinavia found a lower peak age in men [21, 70]; however, this could be attributable to the small numbers of males in these studies (24 males in the study by Nived *et al.* [70] and nine males in the study by Lerang *et al*. [21]).

The majority of studies that compared ethnic differences found Black people to have high incidence and prevalence of SLE, White people to have low and Hispanic and Asian people to have intermediate incidence and prevalence of SLE. However, most of these studies were performed in the USA and Europe. Interestingly, the study of Black Africans in Zimbabwe [9] had a low incidence of SLE. As discussed above, this may have been underestimated. Alternatively, it may be that the incidence and prevalence of SLE is higher in Black populations who have emigrated out of Africa because of differences in gene–environment interactions. This is a hypothesis being explored in the Gullah population in South Carolina compared with people from their ancestral origin in Sierra Leone [108, 109]. Further high-quality epidemiological studies in Africa would also help to address this question. This is challenging in a resource-limited system, where health-care systems are constrained, but could be achieved using the approach used by the WHO-ILAR COPCORD [110].

It is not possible directly to compare the change in incidence and prevalence between studies in the same country that have used different study methods or case definitions; for example, in the UK Nightingale *et al.* [76, 30] used a stricter definition of SLE than Somers *et al.* [31] or Rees *et al.* [1]. The majority of those studies that have looked at the same population using the same methods over time have shown an increasing incidence and prevalence, except for the most recent studies from the UK and the USA, which showed a reduction in incidence. These may be true increases in incidence and prevalence over time, for example, because of an increase in risk factors for SLE and improved survival, or they may be artefactual because of improved diagnosis of people with SLE or better case-ascertainment methods in the study design. Owing to increasing globalization, it is also possibly attributable to net immigration of non-White populations into areas that were previously predominantly White. The recent reductions in incidence in the UK and the USA may therefore reflect changes in environmental risk factors, such as reduced smoking or changes in migration patterns, or perhaps suggest that the risk in later generations of migrants regresses towards the country's mean. It is important to study these temporal changes so that future health services can be planned to meet the needs of the populations.

A potential limitation of this study was that, firstly, for completeness, all eligible studies were included regardless of size or quality. There is therefore a risk of bias affecting the cumulative evidence. In general, earlier studies were less rigorous than more recent studies and there was greater funding of studies in more developed countries. Secondly, as discussed, it is difficult to assess trend over time between studies that have used different methodologies. Future work should consider study design to enable exploration of temporal trends.

Conclusions

In summary, there is wide geographical variation in the reported incidence and prevalence of SLE. North America had the highest reported incidence and prevalence of SLE, Africa had the lowest incidence and Australia the lowest prevalence. The incidence and prevalence of SLE is higher in females compared with males regardless of age or ethnic origin. The incidence and prevalence are age related, and there is a peak incidence and prevalence for both sex. Males have an older peak age of incidence and prevalence compared with females. In general, people of Black ethnicity have the highest incidence and prevalence of SLE worldwide, followed by Asian and then White ethnic groups. There appears to be a trend of increasing prevalence of SLE with time; the trend for incidence is less clear. Further work to address the lack of epidemiological studies of SLE in Africa, for example using the WHO-ILAR COPCORD approach, may further knowledge underpinning ethnic variation in SLE.

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Supplementary data

Supplementary data are available at *Rheumatology* Online.

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