The incidence and prevalence of adult primary Sjögren's Syndrome in New York County

Gordon, Caroline

DOI:
10.1002/acr.23707

Document Version
Peer reviewed version

Citation for published version (Harvard):
Gordon, C 2019, 'The incidence and prevalence of adult primary Sjögren's Syndrome in New York County', *Arthritis Care & Research*, vol. 71, no. 7, pp. 949-960. https://doi.org/10.1002/acr.23707

The Incidence and Prevalence of Adult Primary Sjögren's Syndrome in New York County

Peter M. Izmirly[1]* MD, MSc, Jill P. Buyon[1] MD, Isabella Wan[1] MD, MPH, H. Michael Belmont[1] MD, Sara Sahl[2] MD, MPH, Jane E. Salmon[3] MD, Anca Askanase[4] MD, MPH, Joan M. Bathon[4] MD, Laura Geraldino-Pardilla[4] MD, MSc, Yousaf Ali[5] MD, Ellen M. Ginzler[6] MD, MPH, Chaim Putterman[7] MD, Caroline Gordon[8] MD, Charles G. Helmick[9] MD, and Hilary Parton[10] MPH

[1]Division of Rheumatology, Department of Medicine, New York University School of Medicine, New York, NY; [2]Department of Pediatrics, Harbor-University of California Medical Center Los Angeles, CA; [3]Division of Rheumatology, Department of Medicine, Hospital for Special Surgery, Weill Cornell Medical College, New York, NY; [4]Division of Rheumatology, Department of Medicine Columbia University College of Physicians & Surgeons, New York, NY; [5]Division of Rheumatology, Department of Medicine, Icahn School of Medicine at Mount Sinai, New York, NY; [6]Division of Rheumatology, Department of Medicine, State University of New York Downstate College of Medicine, Brooklyn, NY; [7]Division of Rheumatology, Department of Medicine, Albert Einstein College of Medicine, Bronx, NY; [8]Rheumatology Research Group, Institute of Inflammation and Ageing, College of Medical and Dental Sciences, University of Birmingham, Birmingham, United Kingdom; [9]Arthritis Program, Centers for Disease Control and Prevention, Atlanta, GA; [10]New York City Department of Health and Mental Hygiene, Long Island City, New York

Running Title: Sjögren's Syndrome Incidence and Prevalence in Manhattan

Word Count: 3546

Correspondence to: Peter M. Izmirly, MD

NYU School of Medicine

550 First Avenue, MSB 625

New York, NY 10016

Email: Peter.Izmirly@nyumc.org

Phone: 212-263-5802

Fax: 646-501-5208

The findings and conclusions in this report are those of the authors and do not necessarily represent the official position of the Centers for Disease Control and Prevention.

Conflict of interest: The authors declare no conflict of interest.

Abstract

Objective: Extant epidemiologic data of primary Sjögren's Syndrome (pSS) remains limited, particularly for racial/ethnic populations in the United States (US). The Manhattan Lupus Surveillance Program (MLSP), a population-based retrospective registry of cases with Systemic Lupus Erythematosus and related diseases including pSS in Manhattan, was used to provide estimates of the incidence and prevalence of pSS across major racial/ethnic populations.

Methods: MLSP cases were identified from hospitals, rheumatologists, and population databases. Three case definitions were used for pSS: physician diagnosis, rheumatologist diagnosis, and modified pSS criteria. Rates among Manhattan residents were age-adjusted, and capture-recapture analyses were conducted to assess case under-ascertainment.

Results: By physician diagnosis, age-adjusted overall incidence and prevalence rates of pSS among adult Manhattan residents were 3.5 and 13.1 per 100,000 person-years. Capture-recapture adjustment increased incidence and prevalence rates (4.1 and 14.2). Based on physician diagnosis, incidence and prevalence rates were approximately 6 times higher among women than men ($p<0.01$). Incidence of pSS was statistically higher among non-Latina Asian (10.5) and non-Latina White women (6.2) compared with Latina women (3.2). Incidence was also higher among non-Latina Asian women compared with non-Latina Black women (3.3). Prevalence of pSS did not differ by race/ethnicity. Similar trends were observed when more restrictive case definitions were applied.

Conclusion: Data from the MLSP revealed disparities in pSS incidence and prevalence by sex among Manhattan residents and differences in pSS incidence by race/ethnicity among women. These data also provided epidemiologic estimates for the major racial/ethnic populations in the US.

Significance and Innovations

- This is the first population-based multi-racial/ethnic study in the United States to report on the epidemiology of Sjögren's Syndrome where existing data are sparse in the literature.
- Our study revealed disparities in Sjögren's Syndrome incidence and prevalence by sex among Manhattan residents and differences in incidence by race/ethnicity among women.
- These data also provided epidemiologic estimates for the major racial/ethnic populations in the US.

Sjögren's Syndrome (SS) is a chronic systemic autoimmune disease that manifests as oral and ocular dryness and parotid gland enlargement due to lymphocytic infiltration of exocrine glands, in addition to multi-organ-system extraglandular involvement (1). This syndrome can occur in the absence or presence of other systemic rheumatologic or autoimmune diseases such as Systemic Lupus Erythematosus (SLE), referred to as primary or secondary, respectively. The epidemiology of SS remains limited with few published estimates for the general population and minimal data on multi-racial/ethnic populations in the United States (US) (2, 3).

The Manhattan Lupus Surveillance Program (MLSP) was initiated in 2010 as a collaboration between the New York City Department of Health and Mental Hygiene (DOHMH) and NYU School of Medicine (NYUSoM) (4). The primary goal of the MLSP was to determine incidence and prevalence of SLE among Manhattan residents. To accomplish this, a retrospective population-based registry was established with extensive information obtained on SLE as well as other autoimmune rheumatic diseases including SS. Leveraging this rich data source, we provide incidence and prevalence estimates of primary SS (pSS) during 2007 and 2007-09, respectively, among Manhattan residents across the major racial/ethnic populations (Black, Latino, Asian, White).

Methods

The Manhattan Lupus Surveillance Program

The MLSP is one of five registries funded by the Centers for Disease Control and Prevention (CDC) to provide credible estimates for the incidence and prevalence of SLE (4-8). Details on the MLSP have been previously reported (4). In brief, medical records

were reviewed under the health surveillance exemption to HIPAA privacy rules [45 CFR § 164.512(b)] and as authorized by New York City Charter Sections 556(c)(2) and (d)(2). No cases were contacted for this project. The CDC deemed the various SLE surveillance programs public health practice, which did not require institutional review board (IRB) review, and IRBs at both DOHMH and NYUSoM deemed the MLSP a surveillance activity. When requested, additional IRB applications were completed and submitted to independent case-finding sources. The DOHMH IRB reviewed and approved secondary analyses on a de-identified dataset.

The MLSP surveillance period was January 1, 2007, through December 31, 2009. Manhattan was selected for reasons previously described (4). In 2010, based on US Census data, there were 1,585,873 persons residing in Manhattan (48% non-Latino White, 13% non-Latino Black, 25% Latino, 11% non-Latino Asian) (9).

Case Ascertainment, Data Collection, and Quality Control of Data Entry

The MLSP used the following sources to identify cases: rheumatologists' practices (including pediatric rheumatologists), hospitals, and administrative hospitalization discharge and death registry databases (4). Case finding sources were queried retrospectively, as far back as 2004 when available, for evidence of residence in Manhattan and International Classification of Disease Ninth Revision Clinical Modification (ICD-9CM) billing codes specific for SLE, discoid lupus, and related conditions that may evolve into SLE or have related symptoms including SS. The ICD-9CM codes used to identify cases included 710.0 (SLE), 695.4 (discoid lupus), 710.8 (other specified connective tissue disease), 710.9 (unspecified connective tissue disease), and 710.2 (Sicca syndrome which is used for SS). Charts for every patient

who lived in Manhattan and had one of the respective ICD-9CM codes were fully abstracted and final diagnosis was coded. Abstraction was completed in 90.5% of hospitals and 75.8% of rheumatologists' practices by trained abstractors, all of whom had medical degrees and underwent extensive training and routine quality assurance as previously described (4).

Case Definitions

The MLSP was constructed for surveillance of SLE, and data elements collected focused on two widely used classification schemes for SLE: the American College of Rheumatology (ACR) Criteria (10, 11) and the Systemic Lupus International Collaborating Clinics (SLICC) (12). Additional manifestations commonly associated with SLE – even if not specifically included as a criterion for classification – were also captured, allowing for the potential to identify evidence of SS. Given the overlapping nature of the clinical manifestations of these autoimmune diseases, several but not all of the American-European Consensus Group (AECG) (13) criteria for SS (most recent available at time of data dictionary development) were captured. For criteria regarding the diagnosis of SS not systematically captured, abstractors were trained to take detailed text notes including results of minor salivary gland biopsies and objective results of ocular and oral tests.

Because this analysis focused on pSS, we excluded cases diagnosed with other rheumatologic diseases such as SLE, despite having an additional diagnosis of SS. Also, given the rarity of childhood pSS, we only included cases aged ≥18 years in our analyses (14).

The diagnosis of SS is usually made by a physician familiar with the disease, often but not exclusively by a rheumatologist. Thus, our primary case definition for pSS required documentation of a pSS diagnosis by any physician, and our more conservative secondary case definition required documentation of a pSS diagnosis by a rheumatologist. In the MLSP, few cases met the AECG (13), ACR (15), and the more recent ACR/European League Against Rheumatism (EULAR) criteria for pSS (16). Thus, we developed a third, more restrictive, case definition, slightly modified from the recent ACR/EULAR criteria (16), requiring documentation of all of the following criteria: a) pSS diagnosis by any physician, b) documentation of dry eyes and/or dry mouth, and c) a positive test for anti-SSA antibody.

Statistical Analysis

Incident cases were those aged ≥18 years meeting a pSS case definition, residing in Manhattan, and first diagnosed with pSS from January 1, 2007, through December 31, 2009. Prevalent cases were new or existing cases among those aged ≥18 years meeting a pSS case definition residing in Manhattan from January 1 - December 31, 2007. DOHMH intercensal population estimates for Manhattan were used to calculate denominators (9).

Rates were calculated overall, by sex, and by race/ethnicity per 100,000 person-years and age-adjusted to the US 2000 standard population using 10 year age groups within each racial/ethnic group (17). Although data on race and Latino ethnicity were collected separately during abstraction, cases were assigned to one of five mutually exclusive race/ethnicity categories: non-Latino White, non-Latino Black, non-Latino Asian, Latino, and non-Latino other. Non-Latino cases identified with more than one race were

categorized as other. Chi-square tests or Fisher's exact tests were used to determine if age-adjusted pSS proportions differed by sex and race/ethnicity. When significant differences were found by race/ethnicity, pairwise differences were evaluated using z-tests assuming the Poisson distribution and statistical significance at 0.05, with Bonferroni correction to 0.008.

Capture-recapture analyses were performed (18, 19) to estimate case under-ascertainment; specific methods have been described elsewhere (4). Log-linear models were fit separately for incident and prevalent cases by sex and race/ethnicity for physician-diagnosed cases and by race/ethnicity alone for cases diagnosed by a rheumatologist or meeting the modified case definition due to small numbers.

All analyses were completed using SAS version 9.4 (SAS Institute Inc., Cary, NC, USA) and R version 3.3.0 (R Foundation for Statistical Computing, Vienna, Austria).

Results

Incidence Rates

From 2007-2009, 138 incident cases had a physician diagnosis of pSS and 84 had a rheumatologist diagnosis of pSS. The overall crude and age-adjusted incidence rates for physician-diagnosed cases of pSS were 3.4 (95% confidence interval [CI] 2.9-4.0) and 3.5 (95% CI 2.9-4.1) per 100,000 person-years, respectively (Table 1). The overall crude and age-adjusted incidence rates for rheumatologist-diagnosed cases of pSS were 2.1 (95% CI 1.7-2.6) and 2.1 (95% CI 1.7-2.6) per 100,000 person-years. Age-adjusted rates differed by sex, and were approximately six to seven times higher for women compared with men for both physician- ($p<0.001$) and rheumatologist-diagnosed pSS ($p<0.001$). The incidence of physician-diagnosed pSS differed by race/ethnicity

(p<0.001), with higher rates among Asians (6.2, p=0.002) and Whites (3.8, p=0.006) compared with Latinos (2.0). Incidence of physician-diagnosed pSS was also higher among Asians compared with Blacks (2.2, p=0.005). Similarly, incidence rates also differed by race/ethnicity among women (p<0.01) and were higher among Asian (10.5, p=0.002) and White women (6.2, p=0.007) compared with Latina women (3.2), and among Asian women compared with Black women (3.3, p=0.003). There was no significant difference in age-adjusted incidence of physician-diagnosed pSS by race/ethnicity among men (p=0.859). Incidence of rheumatologist-diagnosed pSS also differed by race/ethnicity overall (p=0.001) and among women (p=0.001), with higher rates among Asian women compared with Latina (p=0.007) and Black women (p=0.006). Capture-recapture adjustment estimated 166.7 incident cases of physician-diagnosed pSS, indicating that 17.2% of cases were missed. Among those missed, 58.9% were White women. The resulting capture-recapture adjusted incidence rate increased to 4.1 per 100,000 person-years (95% CI 2.5-5.8).

The average age (± standard deviation [SD]) at physician diagnosis of pSS was 52.7 (± 18.1) years among women and 58.1 (± 17.3) years among men. The average age (± SD) at diagnosis among incident cases was 56.0 (±19.1) years among Latinos, 54.7 (±18.5) years among Whites, 48.6 (±12.0) years among Blacks, and 47.4 (±18.1) years among Asians.

Among Latino pSS cases, 77.8% of those diagnosed by a physician and 77.8% of those diagnosed by a rheumatologist were also identified as White. Ethnicity information among Latinos was often absent, with two-thirds having no further information available. For those with more detail, ethnicities included Central or South American, Dominican,

Puerto Rican, and Spanish. Among the physician- and rheumatologist-diagnosed incident Asian cases, over one-third had no further data available. Among Asian pSS cases diagnosed by a physician or rheumatologist, >25% had no further classification for Asian ethnicity. Ethnicities among cases with information available included Chinese, Indian or Pakistani, Japanese, and Thai.

Table 2 shows the serologic and clinical manifestations of pSS captured in the MLSP for incident cases of physician- and rheumatologist-diagnosed pSS. Data ascertainment was more complete for cases with a rheumatologist diagnosis. Anti-nuclear antibodies (ANA) and anti-SSA/Ro were the most commonly found serologic manifestations among both physician- and rheumatologist-diagnosed cases. Extraglandular manifestations were present in 62.6% and 65.4% of physician- and rheumatologist-diagnosed cases, with lymphopenia and arthritis being the most common.

Prevalence Rates for pSS

In 2007, a total of 166 cases had a physician diagnosis of pSS and 94 had a rheumatologist diagnosis of pSS. The crude and age-adjusted prevalence of physician-diagnosed pSS overall was 12.4 (95% CI 10.5-14.3) and 13.1 (95% CI 11.1-15.1) per 100,000 person-years (Table 3). The overall crude and age-adjusted prevalence of rheumatologist-diagnosed pSS were lower, at 7.0 (95% CI 5.7-8.6) and 7.3 (95% CI 5.9-8.9) per 100,000 person-years. Age-adjusted rates were approximately six times higher among women compared with men for both physician- ($p<0.001$) and rheumatologist-diagnosed pSS ($p<0.001$). Trends in both physician- and rheumatologist-diagnosed pSS were similar to incidence. The age-adjusted prevalence of physician-diagnosed pSS was 23.8 among White women, 23.7 among Asian women, 16.1 among Black women,

and 15.0 among Latina women. For rheumatologist-diagnosed pSS, Asian women had the highest rate, followed by White, Black, and Latina women. However, there were no significant differences in physician- or rheumatologist-diagnosed prevalence rates by race/ethnicity overall, among women, or among men.

Capture-recapture estimated an additional 24.2 cases of physician-diagnosed pSS, indicating that 12.7% of cases may have been missed. Among cases missed, almost two-thirds (65.3%) were White women. With capture-recapture adjustment, the overall prevalence by physician diagnosis increased to 14.2 per 100,000 person-years (95% CI 12.3-16.1).

The average ages (± SD) among women and men with pSS identified by physician diagnosis were 56.4 (± 17.5) and 60.9 (± 16.9) years respectively. The average age of physician-diagnosed pSS in 2007 was 60.3 (± 16.7) years among Whites, 53.1 (± 20.5) years among Blacks, 52.9 (± 14.8) years among Latinos, and 49.6 (± 19.6) years among Asians.

Among physician- and rheumatologist-diagnosed prevalent Latino cases, more than three-quarters were also identified as White. Information on Latino ethnicity was often absent, with more than two-thirds having no further details. Among Asian pSS cases diagnosed by a physician or rheumatologist, more than a quarter had no further classification for Asian ethnicity.

Table 4 shows the occurrence of relevant serologic and clinical manifestations captured in the MLSP for prevalent physician- and rheumatologist-diagnosed pSS cases. Similar to incident cases, data ascertainment on manifestations was more complete for cases with a rheumatologist diagnosis.

Incidence and Prevalence of pSS using Modified Criteria

Using the modified case definition of pSS (Table 5), incorporating the presence of autoantibodies and documentation of dry eyes and/or dry mouth resulted in an overall age-adjusted incidence rate of 1.1 (95% CI 0.8-1.5) per 100,000 person-years and an overall age-adjusted prevalence rate of 3.3 (95% CI 2.4-4.4) per 100,000 person-years. As with the other case definitions, age-adjusted rates were higher among women compared with men ($p<0.001$). Incidence rates differed by race/ethnicity overall ($p<0.001$), with higher rates among Asians compared with Whites ($p=0.007$) and Latinos ($p=0.005$), and among women ($p<0.001$), with higher rates among Asians compared with Blacks ($p=0.003$). Prevalence of pSS differed by race/ethnicity overall ($p=0.001$) and among women ($p<0.001$) but no significant differences were found by pairwise comparison.

Incident and Prevalent Cases of pSS Meeting Criteria for SLE

Cases with a diagnosis of pSS also met ≥4 of the ACR and/or SLICC criteria for SLE despite not being clinically diagnosed as SLE (Table 6). Depending on the case definition for pSS, 4.3-10.2% of incident cases met the ACR criteria for SLE and 5.8-16.3% met the SLICC criteria. The modified case definition for pSS had the highest percentage of incident cases meeting ACR and SLICC criteria for SLE. There was a higher percentage of prevalent pSS cases meeting SLE criteria (ACR: 6.6-14.9%; SLICC: 14.5-34.0%).

Discussion

Our analysis of the MLSP dataset provides incidence and prevalence rate estimates of pSS among Manhattan residents. These data also provided epidemiologic estimates for

the major racial/ethnic populations in the US. The age-standardized incidence and prevalence of physician-diagnosed pSS in Manhattan were 3.5 (95% CI 2.9-4.1) and 13.1 (95% CI 11.1-15.1) per 100,000 person-years. Capture-recapture adjustment increased incidence and prevalence rates by 17.2% and 12.7%, respectively. By rheumatologist diagnosis, the age-adjusted incidence and prevalence of pSS were 2.1 (95% CI 1.7-2.6) and 7.3 (95% CI 5.9-8.9). Incidence was highest among Asians and Whites, though prevalence did not significantly differ by race/ethnicity, and there were substantial disparities in the prevalence and incidence of pSS among Manhattan residents by sex. This analysis also provides information on serologic and clinical manifestations among pSS cases including data on extraglandular manifestations. In addition, these data reveal that up to a third of prevalent cases diagnosed with pSS also fulfill both ACR and SLICC criteria for SLE, even though they do not carry a diagnosis of SLE, reflecting commonalities in manifestations of the two diseases. Not surprisingly, these data suggest that in clinical practice physicians diagnose patients without formal application of disease criteria.

Previous studies on the epidemiology of SS span decades, come from different regions of the world, and have used varying methods of case identification (2, 3). The few published estimates for the general population reveal annual incidence rates of 6.9–20.1 per 100,000 persons (3) and markedly discrepant prevalence figures ranging from 11.3–3790.1 cases per 100,000 persons (3). Whether these estimates reflect genuine variability between different populations or differences in methodology and study design is unclear.

Existing data suggests the disease is most common in middle-aged women, which is consistent with findings of our analysis (2, 3). In line with these findings, our analyses were restricted to adults aged ≥18 years though it is worth noting that the MLSP did identify pediatric cases of pSS. However, including these pediatric cases into our prevalence and incidence estimates of physician-diagnosed pSS would have decreased our estimates by at least 21% given the small number of cases added to our numerator relative to the person-years added to our denominator.

In a meta-analysis of pSS studies published to date (3), 21 population-based studies were identified, of which only 10 included a review of medical records; the rest were population-based surveys. Six studies (20-25) determined an incidence rate, only one of which was US-based (21); the authors calculated a pooled pSS prevalence rate of 60.8 per 100,000 person-years and an incidence rate of 6.9 per 100,000, both higher than our estimates. However, in line with the findings from our study, the authors found a higher pooled pSS incidence rate among women compared with men (12.3 vs. 1.5). A report limited to European-based studies using the AECG criteria for pSS showed a European prevalence of pSS at 38.95 per 100,000 population (26), and after being updated as a meta-analysis showed a point prevalence of 4.7 per 10,000 population (27).

A recent US-based study of pSS was conducted in Olmsted County, MN, with a mostly White population and reported a population-based prevalence estimate for pSS based on physician diagnosis of 10.3 per 10,000 residents, again higher than our estimate. Even using the conservative AECG definition, the prevalence estimate was still higher than ours at 2.2 (95%CI:1.3–3.1) per 10,000 (28). A separate study of the same

population also provided a higher annual incidence rate of physician-diagnosed pSS at 5.9 per 100,000 population (95%CI:4.4-7.4) (29).

There are virtually no studies that present pSS findings among diverse populations. The meta-analysis (3) presented no information on race or ethnicity other than a few studies done in Taiwan (23-25) which found a pooled incidence rate of 6.6 per 100,000 with significant heterogeneity. One study conducted in the greater Paris area of France reported population-based estimates of pSS prevalence among a multi-racial/multi-ethnic population (2). In line with our findings, prevalence estimates of pSS in this study ranged from 10.0 per 100,000 adults aged ≥15 years to 15.2 per 100,000, depending on the definition used. Prevalence was approximately two times higher for non-Europeans, though incidence and further breakdown on non-European origin was not reported (2). Our study did not find significant differences in prevalence by race/ethnicity but also had a different racial/ethnic makeup. However, we did find significant differences in incidence by race/ethnicity.

There were several limitations regarding the development of the MLSP previously acknowledged (4). These analyses may have underestimated incident and prevalent cases as two hospitals and one quarter of rheumatologists in the catchment area, who practiced in predominantly White neighborhoods, declined to participate. Given that the Veteran's Administration Hospital was one of the hospitals that declined to participate (the other was a cancer specialty hospital), we may have specifically under-identified males diagnosed with pSS. It is also possible that cases were missed if they lived in Manhattan but sought care in other boroughs or a neighboring state. We also did not

include ophthalmologists, otolaryngologists, or primary care practices among our case finding sources.

As previously detailed (4), additional limitations of the MLSP resulted from the tremendous differences across medical systems and abstracting several years after the surveillance period. These limitations could have resulted in abstractors missing information such as results of minor salivary gland biopsies and objective results of ocular and oral tests. This could account for our rates using the modified case definition (16) being considerably lower than those by physician and rheumatologist diagnosis. Another explanation for these lower rates using the modified case definition comes from feedback obtained from our abstractors while in the field. Documentation of salivary gland biopsies or objective evidence of dry eyes (positive Schirmer's test, rose bengal score, or other ocular dye score), dry mouth (positive unstimulated whole salivary flow test, parotid sialography, or salivary scintigraphy), and lip biopsies were rare which limited our planned ability to use the various pSS criteria (13, 15, 16). In addition when biopsies were performed they were not reported in any standardized way (30).

This observation was corroborated in a recent article exploring the prevalence of SS in Olmsted County, MN, where the rates for AECG confirmed SS were considerably lower than rates for physician-diagnosed SS (28). The authors concluded that classification criteria do not accurately reflect the diagnosis of SS in clinical practice in part because the criteria include invasive tests that are rarely performed in routine care (28). Importantly, these criteria sets were not developed for diagnostic use in routine clinical practice, but were designed to capture a more homogeneous patient population for the purpose of research and clinical trials (31).

Additional limitations pertain to assigning race and ethnicity based on administrative and medical records. Though available information did reflect the major ethnic subgroups in Manhattan, specific ethnicity information was missing for most Latino cases and more than one quarter of Asian cases. Categorized broadly, Latino or Asian race encompasses a number of heterogeneous groups and pSS rates among them may differ. Given the already limited number of published studies on pSS among Asians and Latinos, additional work is needed to better describe and understand the epidemiology of pSS among specific ethnic subpopulations.

Despite these limitations, our analysis benefitted from the design and composition of the MLSP (4). First, the MLSP was designed as a population-based registry with a diverse population, which allowed us to estimate rates of pSS among the major racial/ethnic categories. The partnership with the DOHMH allowed us to collect information from a number of case finding sources, which facilitated more complete clinical information on many cases. In addition, we conducted capture-recapture analyses to estimate missed cases. Finally, our abstractors all had medical backgrounds, which helped during training and provided an advantage during extensive review of medical records to identify criteria and manifestations of pSS.

In conclusion, data from a large population-based registry revealed substantial disparities by sex in pSS among Manhattan residents. Differences were also found in the incidence of pSS by race/ethnicity, highlighting higher rates among Asian women which have not been documented previously in the US.

Funding: This work was supported by cooperative agreements between the Centers for Disease Control and Prevention and The New York City Department of Health and

Mental Hygiene [grant number U58/DP002827] and a cooperative agreement between the New York City Department of Health and Mental Hygiene and New York University School of Medicine.

Acknowledgements:

The authors would like to acknowledge Benjamin Wainwright for assistance in preparing the manuscript. The authors wish to thank all of the rheumatologists and their practice managers who participated in the MLSP. We would also like to thank all the administrators in the medical records departments of the hospitals who participated in the MLSP for their assistance obtaining medical records. At the DOHMH, the MLSP would like to acknowledge the contributions of past and current members including Tamira Collins-Bowers, Manasi Joshi, Bonnie Kerker, Maushumi Mavinkurve, Angela Merges, Kyyon Nelson, Viren Shah, Joseph Slade, Lorna Thorpe, Talytha Utley, and Elizabeth Waddell. In addition, the MLSP would like to acknowledge the hard work of their abstractors Drs. Janice McFarlane, Nick Stefanopoulos, Zahira Zahid, Rukayatu Ibrahim, Saleh Massasati, and Simone Shrestha. Finally we would like to acknowledge the support and contribution of the of the principal investigators of the other CDC-funded surveillance sites including Drs. Sam Lim, Cristina Drenkard, Emily Somers, Joe McCune, Maria Dall'Era, and Elizabeth Ferucci.

References

1. Brito-Zeron P, Baldini C, Bootsma H, Bowman SJ, Jonsson R, Mariette X, et al. Sjogren syndrome. Nat Rev Dis Primers. 2016;2:16047.
2. Maldini C, Seror R, Fain O, Dhote R, Amoura Z, De Bandt M, et al. Epidemiology of primary Sjogren's syndrome in a French multiracial/multiethnic area. Arthritis Care Res (Hoboken). 2014;66(3):454-63.
3. Qin B, Wang J, Yang Z, Yang M, Ma N, Huang F, et al. Epidemiology of primary Sjogren's syndrome: a systematic review and meta-analysis. Ann Rheum Dis. 2015;74(11):1983-9.
4. Izmirly PM, Wan I, Sahl S, Buyon JP, Belmont HM, Salmon JE, et al. The Incidence and Prevalence of Systemic Lupus Erythematosus in New York County (Manhattan), New York: The Manhattan Lupus Surveillance Program. Arthritis Rheumatol. 2017;69(10):2006-17.
5. Lim SS, Bayakly AR, Helmick CG, Gordon C, Easley KA, Drenkard C. The incidence and prevalence of systemic lupus erythematosus, 2002-2004: The Georgia Lupus Registry. Arthritis Rheumatol. 2014;66(2):357-68.
6. Somers EC, Marder W, Cagnoli P, Lewis EE, DeGuire P, Gordon C, et al. Population-based incidence and prevalence of systemic lupus erythematosus: the Michigan Lupus Epidemiology and Surveillance program. Arthritis Rheumatol. 2014;66(2):369-78.
7. Ferucci ED, Johnston JM, Gaddy JR, Sumner L, Posever JO, Choromanski TL, et al. Prevalence and incidence of systemic lupus erythematosus in a population-based registry of American Indian and Alaska Native people, 2007-2009. Arthritis Rheumatol. 2014;66(9):2494-502.
8. Dall'Era M, Cisternas MG, Snipes K, Herrinton LJ, Gordon C, Helmick CG. The Incidence and Prevalence of Systemic Lupus Erythematosus in San Francisco County, California: The California Lupus Surveillance Project. Arthritis Rheumatol. 2017;69(10):1996-2005.
9. New York City Department of Health and Mental Hygiene. Epiquery: NYC Interactive Health Data System - NYCDOHMH neighborhood population estimates, modified from the US Census Bureau vintage population estimates, 2007, 2008, 2009. 2016.
10. Hochberg MC. Updating the American College of Rheumatology revised criteria for the classification of systemic lupus erythematosus. Arthritis Rheum. 1997;40(9):1725.
11. Tan EM, Cohen AS, Fries JF, Masi AT, McShane DJ, Rothfield NF, et al. The 1982 revised criteria for the classification of systemic lupus erythematosus. Arthritis Rheum. 1982;25(11):1271-7.
12. Petri M, Orbai AM, Alarcon GS, Gordon C, Merrill JT, Fortin PR, et al. Derivation and validation of the Systemic Lupus International Collaborating Clinics classification criteria for systemic lupus erythematosus. Arthritis Rheum. 2012;64(8):2677-86.
13. Vitali C, Bombardieri S, Jonsson R, Moutsopoulos HM, Alexander EL, Carsons SE, et al. Classification criteria for Sjogren's syndrome: a revised version of the European criteria proposed by the American-European Consensus Group. Ann Rheum Dis. 2002;61(6):554-8.

14. Lieberman SM. Childhood Sjogren syndrome: insights from adults and animal models. Curr Opin Rheumatol. 2013;25(5):651-7.
15. Shiboski SC, Shiboski CH, Criswell L, Baer A, Challacombe S, Lanfranchi H, et al. American College of Rheumatology classification criteria for Sjogren's syndrome: a data-driven, expert consensus approach in the Sjogren's International Collaborative Clinical Alliance cohort. Arthritis Care Res (Hoboken). 2012;64(4):475-87.
16. Shiboski CH, Shiboski SC, Seror R, Criswell LA, Labetoulle M, Lietman TM, et al. 2016 American College of Rheumatology/European League Against Rheumatism classification criteria for primary Sjogren's syndrome: A consensus and data-driven methodology involving three international patient cohorts. Ann Rheum Dis. 2017;76(1):9-16.
17. Klein RJ, Schoenborn CA. Age adjustment using the 2000 projected U.S. population. Healthy People 2010 Stat Notes. 2001(20):1-10.
18. Hook EB, Regal RR. Capture-recapture methods in epidemiology: methods and limitations. Epidemiol Rev. 1995;17(2):243-64.
19. Baillargeon S, Rivest L. Rcapture: Loglinear Models for Capture-Recapture in R. J Stat Softw. 2007;19(5):1-31.
20. Alamanos Y, Tsifetaki N, Voulgari PV, Venetsanopoulou AI, Siozos C, Drosos AA. Epidemiology of primary Sjogren's syndrome in north-west Greece, 1982-2003. Rheumatology (Oxford). 2006;45(2):187-91.
21. Pillemer SR, Matteson EL, Jacobsson LT, Martens PB, Melton LJ, 3rd, O'Fallon WM, et al. Incidence of physician-diagnosed primary Sjogren syndrome in residents of Olmsted County, Minnesota. Mayo Clin Proc. 2001;76(6):593-9.
22. Plesivcnik Novljan M, Rozman B, Hocevar A, Grmek M, Kveder T, Tomsic M. Incidence of primary Sjogren's syndrome in Slovenia. Ann Rheum Dis. 2004;63(7):874-6.
23. See LC, Kuo CF, Chou IJ, Chiou MJ, Yu KH. Sex- and age-specific incidence of autoimmune rheumatic diseases in the Chinese population: a Taiwan population-based study. Semin Arthritis Rheum. 2013;43(3):381-6.
24. Weng MY, Huang YT, Liu MF, Lu TH. Incidence and mortality of treated primary Sjogren's syndrome in Taiwan: a population-based study. J Rheumatol. 2011;38(4):706-8.
25. Yu KH, See LC, Kuo CF, Chou IJ, Chou MJ. Prevalence and incidence in patients with autoimmune rheumatic diseases: a nationwide population-based study in Taiwan. Arthritis Care Res (Hoboken). 2013;65(2):244-50.
26. Cornec D, Chiche L. Is primary Sjogren's syndrome an orphan disease? A critical appraisal of prevalence studies in Europe. Ann Rheum Dis. 2015;74(3):e25.
27. Nocturne G, Seror R, Mariette X, Devauchelle-Pensec V, Saraux A, Chiche L. Primary Sjogren's Syndrome Prevalence: What if Sjogren was Right After All? Comment on the Article by Maciel et al. Arthritis Care Res (Hoboken). 2018;70(6):951-3.
28. Maciel G, Crowson CS, Matteson EL, Cornec D. Prevalence of Primary Sjogren's Syndrome in a US Population-Based Cohort. Arthritis Care Res (Hoboken). 2017;69(10):1612-6.
29. Maciel G, Crowson CS, Matteson EL, Cornec D. Incidence and Mortality of Physician-Diagnosed Primary Sjogren Syndrome: Time Trends Over a 40-Year Period in a Population-Based US Cohort. Mayo Clin Proc. 2017;92(5):734-43.

30. Fisher BA, Jonsson R, Daniels T, Bombardieri M, Brown RM, Morgan P, et al. Standardisation of labial salivary gland histopathology in clinical trials in primary Sjogren's syndrome. Ann Rheum Dis. 2017;76(7):1161-8.
31. Vitali C, Del Papa N. Classification and diagnostic criteria in Sjogren's syndrome: a long-standing and still open controversy. Ann Rheum Dis. 2017;76(12):1953-4.

Table 1: Crude and age-adjusted incidence rates of Sjögren's Syndrome among Manhattan residents aged 18 and older, 2007-2009, overall and by race/ethnicity and sex

	Crude rate (95% CI)	Age-adjusted rate (95% CI)	χ^2 p-value	Capture-Recapture N missed	Capture-Recapture Rate (95% CI)
Primary Sjögren's with Physician Diagnosis					
Total	3.4 (2.9-4.0)	3.5 (2.9-4.1)	<0.001	28.7	4.1 (2.5-5.8)
Male	0.9 (0.5-1.5)	1.0 (0.6-1.5)		4.7	1.2 (0.2-2.1)
Female	5.6 (4.6-6.6)	5.7 (4.7-6.7)		24.0	6.7 (4.3-9.1)
Race/ethnicity			<0.001 [1]		
Non-Latino White	3.7 (3.0-4.7)	3.8 (3.0-4.7)		21.2	4.8 (2.2-7.4)
Non-Latino Black	2.2 (1.1-3.9)	2.2 (1.1-4.0)		1.1	2.4 (1.4-3.4)
Latino	1.9 (1.1-3.0)	2.0 (1.2-3.2)		1.7	2.1 (1.7-2.5)
Non-Latino Asian	5.6 (3.6-8.1)	6.2 (4.0-9.2)		4.3	6.5 (5.2-7.8)
Non-Latino Other	-			0.4	
Race/ethnicity by sex					
Male			0.859		
Non-Latino White	1.1 (0.6-2.0)	1.1 (0.5-2.0)		4.3	1.6 (0.1-3.1)
Non-Latino Black	0.9 (0.1-3.2)	0.9 (0.1-3.2)		0.2	1.0 (0.3-1.7)
Latino	0.5 (0.1-1.7)	0.5 (0.1-1.7)		0.2	0.5 (0.1-0.9)
Non-Latino Asian	0.5 (0.0-2.7)	0.6 (0.0-3.6)		0.0	0.5 (0.4-0.6)
Female			<0.001 [2]		
Non-Latina White	6.1 (4.7-7.8)	6.2 (4.7-7.9)		16.9	7.7 (4.1-11.3)
Non-Latina Black	3.2 (1.5-6.1)	3.3 (1.5-6.3)		0.9	3.5 (2.3-4.8)
Latina	3.2 (1.8-5.1)	3.2 (1.8-5.2)		1.5	3.5 (3.1-3.8)
Non-Latina Asian	9.5 (6.2-14.0)	10.5 (6.6-15.7)		4.3	11.1 (8.9-13.4)
Primary Sjögren's with Rheumatologist Diagnosis					
Total	2.1 (1.7-2.6)	2.1 (1.7-2.6)	<0.001	34.0	2.9 (1.1-4.8)
Male	0.5 (0.2-0.9)	0.5 (0.2-0.9)			
Female	3.5 (2.7-4.4)	3.5 (2.7-4.4)			
Race/ethnicity			0.001 [1]		
Non-Latino White	2.4 (1.8-3.2)	2.3 (1.7-3.1)		13.2	3.0 (2.1-4.0)
Non-Latino Black	1.2 (0.4-2.6)	1.2 (0.5-2.7)		9.0	2.9 (-1.8-7.7)
Latino	1.0 (0.4-1.8)	0.9 (0.4-1.8)		0.6	1.0 (0.6-1.5)
Non-Latino Asian	3.8 (2.3-6.1)	4.1 (2.3-6.5)		10.7	6.1 (1.0-11.2)

Non-Latino Other	-			0.5
Race/ethnicity by sex				
Male			0.524	
Non-Latino White	0.6 (0.2-1.3)	0.5 (0.2-1.2)		
Non-Latino Black	0.9 (0.1-3.2)	0.9 (0.1-3.2)		
Latino	-			
Non-Latino Asian	0.5 (0.0-2.7)	0.6 (0.0-3.6)		
Female			0.001 [3]	
Non-Latina White	4.0 (2.9-5.4)	3.8 (2.8-5.2)		
Non-Latina Black	1.4 (0.4-3.6)	1.6 (0.4-4.1)		
Latina	1.8 (0.8-3.4)	1.7 (0.8-3.3)		
Non-Latina Asian	6.5 (3.8-10.3)	6.7 (3.8-11.1)		

Rates are per 100,000 Manhattan residents. Denominator data is based on 2007-2009 intercensal population estimates from the NYC DOHMH Bureau of Epi Services (2000-2014 files).
Data are age adjusted to the US2000 Standard Population.
Cases were assigned to one of five mutually exclusive race/ethnicity categories: non-Latino white, non-Latino black, non-Latino Asian, Latino, and non-Latino other. Non-Latino cases identified with more than one race were categorized as non-Latino other.
For capture-recapture analyses, log-linear models were fit separately for by sex and race/ethnicity for physician diagnosed cases and by sex alone for cases diagnosed by rheumatologist or meeting the modified case definition.
[1] Latinos differed from non-Latino Whites and non-Latino Asians. Non-Latino Asians also differed from non-Latino Blacks.
[2] Latinas differed from non-Latina Whites and non-Latina Asians. Non-Latina Asians also differed from non-Latina Blacks.
[3] Latinas differed from non-Latina Whites and non-Latina Asians. Non-Latina Blacks also differed from non-Latina Whites and non-Latina Asians.

Table 2: Frequency of specific manifestations among incident Sjögren's Syndrome cases among NYC Manhattan residents aged 18 and older, 2007-2009

	Primary Sjogren's with Physician Diagnosis			Primary Sjogren's with Rheumatologist Diagnosis			Primary Sjogren's - modified definition		
	Number available	Positive N	%	Number available	Positive N	%	Number available	Positive N	%
Overall N	138			84			45		
Glandular/serologies									
Sicca symptoms	122	91	74.6%	82	72	87.8%	45	45	100.0%
Anti-SSA/Ro	96	63	65.6%	78	52	66.7%	45	45	100.0%
Anti-SSB/La	90	37	41.1%	75	30	40.0%	42	23	55.0%
Anti-SSA/Ro and Anti-SSB/La	90	33	36.7%	75	26	34.7%	42	23	55.0%
ANA	90	71	78.9%	72	61	84.7%	39	37	95.0%
ANA titer >1:320	45	30	66.7%	39	25	64.1%	25	16	64.0%
Rheumatoid factor	67	27	40.3%	53	22	41.5%	31	17	55.0%
Extraglandular									
Arthritis	137	29	21.2%	83	19	22.9%	44	10	23.0%
Photo sensitivity	138	7	5.1%	84	7	8.3%	45	4	9.0%
Lymphopenia	124	64	51.6%	79	39	49.4%	42	22	52.0%
Interstitial lung disease	138	4	2.9%	84	2	2.4%	45	2	4.0%
Pneumonitis	138	1	0.7%	84	1	1.2%	45	1	2.0%
Transverse myelitis	138	0	0.0%	84	0	0.0%	45	0	0.0%
Low complements	138	5	3.6%	84	5	6.0%	45	5	11.0%
Raynaud's	138	13	9.4%	84	9	10.7%	45	3	7.0%
Cutaneous vasculitis	138	0	0.0%	84	0	0.0%	45	0	0.0%
Cranial or peripheral neuropathy	137	10	7.3%	84	5	6.0%	45	2	4.0%
Myositis	137	0	0.0%	83	0	0.0%	44	0	0.0%

Table 3: Crude and age-adjusted prevalence rates of Sjögren's Syndrome among Manhattan residents aged 18 and older, 2007, overall and by race/ethnicity and sex

	Crude rate (95% CI)	Age-adjusted rate (95% CI)	χ^2 p-value	Capture-Recapture N missed	Capture-Recapture Rate (95% CI)
Primary Sjögren's with Physician Diagnosis					
Total	12.4 (10.5-14.3)	13.1 (11.1-15.1)	<0.001	24.2	14.2 (12.3-16.1)
Male	3.1 (1.9-4.8)	3.5 (2.1-5.5)		0.1	3.1 (3.0-3.2)
Female	20.5 (17.2-23.8)	21.1 (17.6-24.5)		24.1	23.9 (20.3-27.4)
Race/ethnicity			0.099		
Non-Latino White	13.9 (11.3-17.0)	14.6 (11.8-17.9)		15.9	16.3 (14.4-18.1)
Non-Latino Black	9.4 (5.4-15.2)	9.4 (5.4-15.4)		0.5	9.6 (8.4-10.9)
Latino	8.6 (5.7-12.6)	9.1 (6.0-13.2)		0.6	8.8 (8.1-9.5)
Non-Latino Asian	13.1 (8.0-20.2)	14.3 (8.5-22.5)		6.1	17.1 (11.6-22.6)
Non-Latino Other				1.1	
Race/ethnicity by sex					
Male			0.638		
Non-Latino White	4.0 (2.1-6.9)	4.3 (2.3-7.4)		0.1	4.0 (3.8-4.3)
Non-Latino Black	1.3 (0.0-7.3)	1.7 (0.0-9.4)		0.0	1.3 (1.3-1.3)
Latino	1.4 (0.2-5.0)	1.5 (0.2-5.4)		0.0	1.4 (1.4-1.4)
Non-Latino Asian	1.5 (0.0-8.3)	2.2 (0.1-12.5)		0.0	1.5 (1.5-1.5)
Female			0.153		
Non-Latina White	22.9 (18.2-28.4)	23.8 (18.9-29.6)		15.8	27.3 (24.0-30.7)
Non-Latina Black	15.8 (8.9-26.1)	16.1 (8.9-26.7)		0.5	16.3 (14.1-18.6)
Latina	14.9 (9.6-21.9)	15.0 (9.7-22.2)		0.6	15.2 (13.9-16.5)
Non-Latina Asian	22.2 (13.4-34.6)	23.7 (13.9-37.7)		6.1	29.3 (19.5-39.1)
Primary Sjögren's with Rheumatologist Diagnosis					
Total	7.0 (5.7-8.6)	7.3 (5.9-8.9)	<0.001	27.3	9.1 (6.2-11.9)
Male	1.6 (0.8-3.0)	1.8 (0.9-3.4)			
Female	11.7 (9.3-14.5)	11.9 (9.5-14.8)			
Race/ethnicity			0.399		
Non-Latino White	7.2 (5.3-9.5)	7.5 (5.5-10.0)		17.5	9.7 (7.5-12.0)
Non-Latino Black	5.3 (2.4-10.0)	5.6 (2.5-10.6)		1.3	6.0 (4.2-7.9)
Latino	5.4 (3.2-8.7)	5.6 (3.2-9.0)		2.0	6.1 (5.0-7.2)
Non-Latino Asian	9.2 (5.0-15.4)	9.7 (5.1-16.6)		3.6	11.5 (5.5-17.5)
Non-Latino Other				2.9	

Race/ethnicity by sex			
Male			0.703
Non-Latino White	2.2 (0.9-4.5)	2.4 (0.9-4.9)	
Non-Latino Black	-		
Latino	0.7 (0.0-3.9)	0.8 (0.0-4.3)	
Non-Latino Asian	1.5 (0.0-8.3)	2.2 (0.1-12.5)	
Female			0.490
Non-Latina White	11.7 (8.5-15.9)	12.1 (8.7-16.5)	
Non-Latina Black	9.5 (4.3-18.0)	10.4 (4.7-19.8)	
Latina	9.5 (5.4-15.4)	9.4 (5.4-15.3)	
Non-Latina Asian	15.2 (8.1-25.9)	15.6 (8.0-27.3)	

Rates are per 100,000 Manhattan residents. Denominator data is based on 2007-2009 intercensal population estimates from the NYC DOHMH Bureau of Epi Services (2000-2014 files)
Data are age adjusted to the US2000 Standard Population.
Cases were assigned to one of five mutually exclusive race/ethnicity categories: non-Latino white, non-Latino black, non-Latino Asian, Latino, and non-Latino other. Non-Latino cases identified with more than one race were categorized as non-Latino other.
For capture-recapture analyses, log-linear models were fit separately for by sex and race/ethnicity for physician diagnosed cases and by sex alone for cases diagnosed by rheumatologist or meeting the modified case definition.

Table 4: Frequency of specific manifestations among prevalent Sjögren's Syndrome cases among NYC Manhattan residents aged 18 and older, 2007

	Primary Sjögren's with Physician Diagnosis			Primary Sjögren's with Rheumatologist Diagnosis			Primary Sjögren's - modified definition		
	Number available	Positive N	%	Number available	Positive N	%	Number available	Positive N	%
Overall N	166			94			47		
Glandular/serologies									
Sicca symptoms	152	110	72.4%	91	81	89.0%	44	44	100.0%
Anti-SSA/Ro	102	60	58.8%	77	48	62.3%	44	44	100.0%
Anti-SSB/La	100	47	47.0%	77	37	48.1%	44	32	73.0%
Anti-SSA/Ro and Anti-SSB/La	100	44	44.0%	77	34	44.2%	44	32	73.0%
ANA	106	72	67.9%	81	58	71.6%	44	38	86.0%
ANA titer >1:320	56	32	57.1%	46	27	58.7%	31	21	68.0%
Rheumatoid factor	82	42	51.2%	64	36	56.3%	35	26	74.0%
Extraglandular									
Arthritis	166	37	22.3%	94	26	27.7%	44	16	36.0%
Photo sensitivity	166	5	3.0%	94	5	5.3%	44	3	7.0%
Lymphopenia	149	103	69.1%	87	61	70.1%	44	35	80.0%
Interstitial lung disease	166	10	6.0%	94	5	5.3%	44	3	7.0%
Pneumonitis	166	3	1.8%	94	1	1.1%	44	1	2.0%
Transverse myelitis	166	0	0.0%	94	0	0.0%	44	0	0.0%
Low complements	166	10	6.0%	94	10	10.6%	44	7	16.0%
Raynaud's	166	16	9.6%	94	14	14.9%	44	5	11.0%
Cutaneous vasculitis	166	6	3.6%	94	3	3.2%	44	2	5.0%
Cranial or peripheral neuropathy	165	17	10.3%	94	12	12.8%	44	5	11.0%
Myositis	166	2	1.2%	94	1	1.1%	44	1	2.0%

Table 5: Crude and age-adjusted rates of Primary Sjögren's Syndrome by Modified Definition among Manhattan residents aged 18 and older, overall and by race/ethnicity and sex

				Capture-Recapture	
	Crude rate (95% CI)	Age-adjusted rate (95% CI)	χ^2 p-value	N missed	Rate (95% CI)
Incidence, 2007-2009					
Total	1.1 (0.8-1.5)	1.1 (0.8-1.5)	<0.001	14.1	1.5 (0.3-2.6)
Male	0.2 (0.1-0.6)	0.2 (0.1-0.5)			
Female	1.9 (1.4-2.6)	1.8 (1.3-2.5)			
Race/ethnicity			<0.001 [1]		
Non-Latino White	0.9 (0.6-1.4)	0.9 (0.5-1.3)		2.0	1.0 (0.6-1.4)
Non-Latino Black	0.8 (0.2-2.0)	0.8 (0.2-2.1)		2.4	1.3 (-1.1-3.6)
Latino	0.7 (0.3-1.5)	0.7 (0.3-1.5)		1.2	0.9 (0.2-1.5)
Non-Latino Asian	3.0 (1.6-5.0)	3.3 (1.6-5.2)		8.2	4.7 (0.8-8.7)
Non-Latino Other	-			0.3	
Race/ethnicity by sex					
Male			0.004		
Non-Latino White	0.1 (0.0-0.6)	0.1 (0.0-0.5)			
Non-Latino Black	0.9 (0.1-3.2)	0.9 (0.1-3.2)			
Latino	-				
Non-Latino Asian	0.5 (0.0-2.7)	0.6 (0.0-3.6)			
Female			<0.001 [2]		
Non-Latina White	1.7 (1.0-2.6)	1.5 (0.9-2.3)			
Non-Latina Black	0.7 (0.1-2.6)	0.7 (0.1-2.7)			
Latina	1.4 (0.6-2.8)	1.4 (0.5-2.8)			
Non-Latina Asian	4.9 (2.6-8.5)	4.9 (2.5-8.6)			
Prevalence, 2007					
Total	3.3 (2.4-4.4)	3.3 (2.4-4.4)	<0.001	18.6	4.7 (1.4-8.0)
Male	0.5 (0.1-1.4)	0.5 (0.1-1.4)			
Female	5.7 (4.1-7.8)	5.7 (4.1-7.8)			
Race/ethnicity			0.001		
Non-Latino White	2.8 (1.7-4.4)	2.8 (1.6-4.3)		12.3	4.6 (0.6-8.6)
Non-Latino Black	3.5 (1.3-7.6)	3.6 (1.3-7.8)		0.8	4.0 (2.6-5.3)

Latino	3.2 (1.5-5.9)	3.1 (1.5-5.7)		2.5	4.0 (2.4-5.6)
Non-Latino Asian	5.2 (2.3-10.3)	5.2 (2.1-10.5)		2.1	6.6 (3.0-10.2)
Non-Latino Other	-			0.9	
Race/ethnicity by sex					
Male					
Non-Latino White	0.9 (0.2-2.7)	0.9 (0.2-2.8)			
Non-Latino Black	-				
Latino	-				
Non-Latino Asian					
Female			0.001		
Non-Latina White	4.5 (2.6-7.3)	4.8 (2.5-7.2)			
Non-Latina Black	6.3 (2.3-13.8)	6.6 (2.4-14.5)			
Latina	5.9 (2.9-10.9)	5.8 (2.7-10.7)			
Non-Latina Asian	9.3 (4.0-18.4)	9.4 (3.9-19.1)			

Rates are per 100,000 Manhattan residents. Denominator data is based on 2007-2009 intercensal population estimates from the NYC DOHMH Bureau of Epi Services (2000-2014 files).
Data are age adjusted to the US2000 Standard Population.
Cases were assigned to one of five mutually exclusive race/ethnicity categories: non-Latino white, non-Latino black, non-Latino Asian, Latino, and non-Latino other. Non-Latino cases identified with more than one race were categorized as non-Latino other.
For capture-recapture analyses, log-linear models were fit separately for by sex and race/ethnicity for physician diagnosed cases and by sex alone for cases diagnosed by rheumatologist or meeting the modified case definition.
[1] Non-Latino Asians differed from non-Latino Whites and Latinos.
[2] Non-Latina Asians differed from non-Latina Blacks.

Table 6: Frequency of specific manifestations among primary Sjögren's Syndrome cases among NYC Manhattan residents aged 18 and older

	Incident cases, 2007-2009					
	Physician Diagnosis		Rheumatologist Diagnosis		Modified Definition	
	N	% positive	N	% positive	N	% positive
Overall N	138		84		49	
Meet ACR SLE criteria		4.3%		7.1%		10.2%
Meet SLICC SLE criteria		5.8%		9.5%		16.3%
	Prevalent cases, 2007					
	Physician Diagnosis		Rheumatologist Diagnosis		Modified Definition	
	N	% positive	N	% positive	N	% positive
Overall N	166		94		47	
Meet ACR SLE criteria		6.6%		10.6%		14.9%
Meet SLICC SLE criteria		14.5%		23.4%		34.0%