Systemic Sclerosis and Associated Interstitial Lung Disease in Ontario, Canada: An Examination of Prevalence and Survival Over 10 Years

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ABSTRACT. **Objective.** Systemic sclerosis (SSc) is a rare autoimmune disease. Pulmonary complications of SSc are some of the leading causes of morbidity and mortality. The objective of this study was to determine prevalence and survival estimates of SSc and SSc with interstitial lung disease (SSc-ILD) in the Canadian province of Ontario using administrative data over 10 years.

Methods. Using International Classification of Diseases, 10th revision codes adapted for Canada (ICD-10-CA), adult patients diagnosed with SSc and SSc-ILD between April 1, 2008, and March 31, 2018, were identified from the National Ambulatory Care Reporting System and the Discharge Abstract Database administrative databases. SSc was identified first, and ILD was included if presence occurred after SSc diagnosis. Prevalence estimates were determined for both SSc and SSc-ILD. For survival rates, Kaplan-Meier survival curves were generated.

Results. At the start of the 2017/18 fiscal year (final year of the cohort), there were 2114 prevalent SSc cases for a cumulative prevalence of 19.1 per 100,000 persons, as well as 257 prevalent cases of SSc-ILD that generated a prevalence of 2.3 cases per 100,000 persons. Mean ages were 57 and 58 years with 84% and 80% females for patients with SSc and SSc-ILD, respectively. One-, 5-, and 10-year survival rates were 85.0%, 64.5%, and 44.9% for the SSc group and 77.1%, 44.4%, and 22.0% for the SSc-ILD group, respectively.

Conclusion. To our knowledge, this study provides the first population-based estimates of SSc and SSc-ILD in Canada for prevalence and survival. Results confirm that the prevalence estimates of SSc-ILD fall within the Canadian threshold for rare disease. It also demonstrates the poor survival in SSc, especially when ILD is also present.

Key Indexing Terms: interstitial lung disease, prevalence, survival, systemic sclerosis

Systemic sclerosis (SSc) is a rare and complex chronic connective tissue disease, characterized by immune dysregulation, microvascular damage, and progressive fibrosis. Often there is severe organ involvement and increased risk of complications and rapid decline, leading to an unpredictable clinical course and a high disease burden. Estimates of the prevalence of SSc vary greatly depending on the methodology used for case ascertainment.[1] Diagnosis of SSc can be difficult due to its clinical heterogeneity and variety of organ manifestations.[2] In the European Union, SSc prevalence estimates range from 7.2 per 100,000 persons in Norway for the year 2009, to 33.9 per 100,000 persons in Italy in 2004.[3,4] In North America, one prevalence estimate of SSc in 2008 was 18.4 per 100,000 persons.[5] Thompson, et al[6] examined a small population in Southwestern Ontario and estimated the prevalence to be 28 per 100,000 persons.

The peak age of onset for SSc is between 40 and 60 years, with women being 4 times more likely to develop the disease than men.[7] Results from studies conducted in 1999–2009 reported that the mean age of patients with SSc at diagnosis was 51.6 (± 13.7) years in the UK and 47 years in Norway, respectively.[3,8] Similarly, a population-based cohort study conducted in the United States reported a median age at diagnosis of 49.1 years.[9]

Interstitial lung disease (ILD) is a common manifestation of SSc, associated with increased morbidity and mortality; however, few published estimates of the prevalence and survival of SSc-ILD are available. Literature suggests that most patients with SSc have some evidence of lung disease, including ILD

and pulmonary hypertension.[8] ILD is among the leading causes of death related to SSc. Patients with SSc-ILD have a median survival of 5 to 8 years.[10,11] Estimates of SSc-ILD prevalence vary greatly: studies in Europe and North America show that 18.8–60.0% and 15.0–52.3%, respectively, of patients with SSc develop SSc-ILD.[9,12,13,14] The most recent Canadian estimates of SSc-ILD are based on the Canadian Scleroderma Research Group Registry (CSRG).[14] Steele, *et al*[14] examined a sample of 1168 adult patients with SSc from the CSRG registry across 15 centers in Canada. SSc diagnosis was first confirmed by a rheumatologist in adult patients. To estimate the presence of ILD, an algorithm using clinical measures was created to define SSc-ILD, which determined the prevalence of SSc-ILD to be 52%.[14] However, the findings represent a highly selected SSc patient population in the registry and referred only to tertiary care centers, potentially making the results subject to referral bias. A systematic literature review of SSc suggested that most severe organ complications, including ILD, occur at a frequency of 15%.[15]

Diagnoses of SSc and subsequent SSc-ILD occur predominantly in women.[16] In another systematic review, the ratio of females to males diagnosed in Europe ranged from 4:1 to 12:1, and in North America, the ratio was about 8:1. They also found that the mean age at diagnosis of SSc-ILD was higher in older patients (61.8 yrs). Similarly, a US study reported the mean age at SSc-ILD diagnosis as 54 years.[17]

Ontario is the largest and most populous province in Canada, with a population of 14.6 million.[18] All citizens have universal health care with respect to access to physician visits and hospitalizations. This large population with single-payer reimbursement for health care makes it an excellent resource to determine the prevalence of rare diseases.

To date, no published study has generated population-based estimates of prevalence and survival rates for SSc-ILD in Canada. The primary objective of this study is to determine prevalence and survival estimates of SSc and SSc-ILD in Ontario, Canada.

METHODS

Study design. A noninterventional, retrospective population-based study was conducted with data from April 1, 2008, to March 31, 2018, in order to generate prevalence estimates using administrative hospital and ambulatory care data in Ontario. Patients were first identified as having SSc using the Canadian Institute for Health Information (CIHI) National Ambulatory Care Reporting System (NACRS) and the Discharge Abstract Database (DAD), using diagnosis codes from the International Classification of Diseases, 10th revision, adapted for Canada (ICD-10-CA): M34.X codes indicating SSc disease (M34, M34.0, M34.1, M34.2, M34.8, M34.9), followed by the ICD codes for ILD, with any of the following indicating pulmonary fibrosis: J84.1, J84.8, J84.9, or J99.1. A validation study of the CIHI-DAD conducted by the Institute for Clinical Evaluative Sciences (ICES) found that coding of data elements occurs with a high degree of accuracy. The most responsible diagnosis tends to be coded well.[19] The majority of ICD codes for SSc have been validated for use in hospital databases such as DAD and NACRS, with the exception of M34.2 (SSc induced by drugs and chemicals). We expanded on the validated codes M34.0, M34.1, M34.8, and M34.9 to include M34.2 as a plausible SSc diagnosis based on clinical input.[20] The ILD codes selected have been established for idiopathic pulmonary fibrosis (IPF), which is a subtype of ILD. Generally,

IPF is classified under the J84 code, and as a result we included all J84 codes available based on the ICD-10-CA (i.e., J84.1, J84.8, and J84.9), which have been used in previous IPF studies.[21] The definition was expanded to include J99.1 (respiratory disorders in other connective tissue disorders) based on clinical input to suggest that some patients with SSc may receive this diagnosis for ILD. However, there is no consensus on a validated algorithm to define ILD in administrative data.[22]

In the primary analysis, the index date was defined as the date of SSc diagnosis for both patients with SSc and those with SSc-ILD, with the ILD diagnosis occurring after the SSc diagnosis. Demographic and epidemiological information was examined. The study population included all living Ontario adult residents (age ≥ 18 yrs) with valid provincial health coverage who had a date of last contact with the healthcare system in Ontario within the past 7 years (from the first day of each fiscal year) during this study period. Patients were excluded if they had an ILD diagnosis prior to the SSc diagnosis or if they had missing or incomplete information at the index date. Figure 1 shows the flow chart for included cases.

Data sources. ICES collects population-level health information in order to generate real-world data available for research through a number of datasets. For this study, data from April 1, 2008, to March 31, 2018, were used to identify prevalent patients with SSc and SSc-ILD from 2 databases: the DAD and NACRS. The DAD includes administrative, clinical, and demographic information on acute inpatient hospital discharges (including deaths, sign-outs, and transfers). The NACRS database consists of emergency department visits and hospital-based ambulatory care such as day surgery and clinic visits.

Outcomes. The primary outcome in this study was to estimate the crude prevalence of SSc and SSc-ILD per 100,000 persons. Secondary outcomes included identifying survival estimates for both patients with SSc and those with SSc-ILD over the 10-year study period, as well as demographic and clinical characteristics.

Statistical analyses. The results were reported at an aggregate level and tabulated, and all analyses were conducted by an ICES analyst, using the index date (i.e., the date of SSc diagnosis) as the primary analysis. Descriptive statistics were used to evaluate the prevalence of the study cohorts. Patient and demographic characteristics were summarized for patients in the SSc and SSc-ILD cohorts by number and percentage for categorical variables (e.g., sex) and by mean and SD for continuous variables (e.g., age). Cells with a size of ≤ 5 were suppressed for privacy reasons and to reduce the chance of reidentification.[23]

Prevalence estimate calculation. Prevalence was calculated by dividing the number of patients with an SSc or SSc-ILD diagnosis by the total population drawn from the start of the 2017–2018 fiscal year to generate a cumulative prevalence estimate over 10 years. The SSc and SSc-ILD patient counts were generated at the start of the respective fiscal year. Prevalence estimates were stratified by factors such as age and sex.

Survival analysis. Kaplan-Meier survival curves were generated for both the SSc and SSc-ILD cohorts, starting at date of diagnosis and continuing to death (for those with the known date of death). Otherwise, patients still alive were censored.

Sensitivity analysis. A sensitivity analysis was conducted by changing the index date from SSc-ILD to ILD diagnosis date (any of the 4 "J" diagnosis codes, as outlined in Supplementary Data 1, available with the online version of this article). This served as a narrowing of the definition of SSc-ILD. Prevalence estimates of SSc-ILD generated using this index date were determined.

Patient and public involvement. Due to the administrative nature of this study, all aspects of this research were conducted without patient involvement.

Ethics review. This study was approved by Veritas Independent Review Board (IRB #16314).
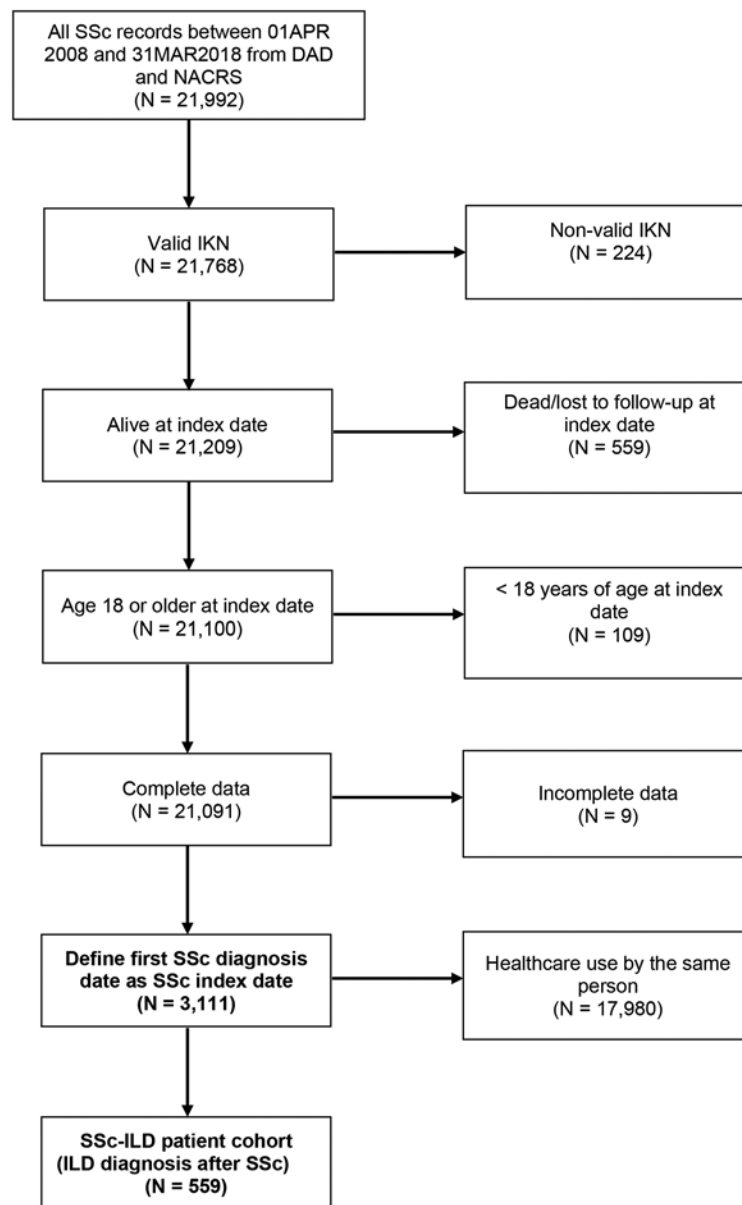
Figure 1. Flow chart of SSc and SSc-ILD included cases. DAD: Discharge Abstract Database; ILD: interstitial lung disease; IKN: Institute for Clinical Evaluative Sciences key number; NACRS: National Ambulatory Care Reporting System; SSc: systemic sclerosis.

RESULTS

Prevalent population baseline characteristics. For the primary analysis over the 10-year study period (2008/9–2017/18), there were 2114 prevalent cases of SSc, and 257 prevalent cases of SSc-ILD identified. To be considered a prevalent case, patients had to be alive and have a positive diagnosis of SSc and/or SSc-ILD during that particular year. Table 1 outlines the baseline characteristics for the prevalent SSc and SSc-ILD cohorts. The mean age at the index date for patients with SSc was 57.4 years, and 84.2% were female. For patients with SSc-ILD, the mean age at index date was 57.9 years, and 80.2% were female.

The sensitivity analysis also identified 257 prevalent SSc-ILD patients over the 10-year study period using the diagnosis date of

ILD (to 2017/18). The mean age at ILD index date was slightly higher at 59.9 years, and 80.2% were female. Baseline characteristics of the sensitivity analysis can be found in Supplementary Table 1 (available with the online version of this article).

Table 2 shows the number of prevalent cases (by diagnosis year and cumulative) for both the SSc and SSc-ILD groups by fiscal year (2008/9–2017/18) of the study period. Table 3 shows the overall prevalence of SSc and SSc-ILD for the primary analysis by age group and sex in both groups. Of the 2114 prevalent SSc cases, the cumulative prevalence rate at the start of fiscal year 2017/18 was found to be 19.1 per 100,000 persons. Patients aged 65+ had a higher cumulative prevalence (29.4 per 100,000 persons), and females had a higher overall prevalence (31.2

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Table 1. Baseline characteristics for prevalent patients with SSc and SSc-ILD over the study period.

Patients with SSc

Year	2008/9 n = 497	2009/10 n = 823	2010/11 n = 1073	2011/12 n = 1239	2012/13 n = 1435	2013/14 n = 1590	2014/15 n = 1749	2015/16 n = 1909	2016/17 n = 2007	2017/18 n = 2114
Age at index date, yrs										
Mean ± SD	60.0 ± 14.4	59.6 ± 14.6	59.3 ± 14.4	58.6 ± 14.2	58.2 ± 14.3	58.2 ± 14.2	58.0 ± 14.2	57.8 ± 14.4	57.6 ± 14.4	57.4 ± 14.3
Median (IQR)	60 (51–72)	60 (50–71)	60 (50–70)	59 (49–69)	58 (49–69)	59 (49–68)	58 (49–68)	58 (48–68)	58 (48–68)	58 (48–67)
Age group, yrs, n (%)										
18–29	10 (2.0)	18 (2.2)	23 (2.2)	32 (2.6)	48 (3.3)	53 (3.3)	57 (3.3)	64 (3.4)	69 (3.4)	72 (3.4)
30–50	114 (22.9)	199 (24.2)	263 (24.5)	309 (24.9)	354 (24.7)	390 (24.5)	441 (25.2)	493 (25.8)	523 (26.1)	555 (26.3)
51–64	180 (36.2)	299 (36.3)	400 (37.3)	471 (38.0)	533 (37.1)	584 (36.7)	645 (36.9)	702 (36.8)	745 (37.1)	799 (37.8)
65+	193 (38.8)	307 (37.3)	387 (36.1)	427 (34.5)	500 (34.8)	563 (35.4)	606 (34.7)	650 (34.1)	670 (33.4)	688 (32.5)
Sex, n (%)										
Female	412 (82.9)	698 (84.8)	914 (85.2)	1060 (85.5)	1215 (84.7)	1353 (85.1)	1492 (85.3)	1619 (84.8)	1698 (84.6)	1780 (84.2)
Male	85 (17.1)	125 (15.2)	159 (14.8)	179 (14.5)	220 (15.3)	237 (14.9)	257 (14.7)	290 (15.2)	309 (15.4)	334 (15.8)

Patients With SSc–ILD

Year	2008/9 n = 118	2009/10 n = 178	2010/11 n = 227	2011/12 n = 262	2012/13 n = 282	2013/14 n = 288	2014/15 n = 289	2015/16 n = 296	2016/17 n = 281	2017/18 n = 257
Time to ILD after SSc diagnosis, d										
Mean ± SD	964.5 ± 981.5	974.4 ± 961.9	987.3 ± 910.5	971.8 ± 886.9	975.7 ± 870.8	958.3 ± 861.8	935.0 ± 866.2	865.0 ± 855.3	798.7 ± 846.8	732.2 ± 840.1
Median (IQR)	629.5 (107–1632)	735.5 (118–1632)	810 (176–1609)	844.5 (181–1572)	848.5 (206–1572)	801.5 (222.5–1545)	771 (167–1494)	611 (144.5–1348)	511 (100–1246)	390 (68–1129)
Age at index date, yrs										
Mean ± SD	59.1 ± 13.3	57.8 ± 13.1	57.8 ± 12.7	58.4 ± 12.5	59.0 ± 12.3	58.9 ± 12.3	58.4 ± 11.9	58.5 ± 12.4	58.1 ± 12.2	57.9 ± 12.2
Median (IQR)	59 (52–69)	57 (50–67)	58 (49–67)	59 (50–67)	60 (51–68)	60 (51–68)	59 (51–67)	59 (50–67.5)	59 (50–67)	59 (50–66)
Age group, yrs, n (%)										
18–29	1–5[a]	1–5[a]	1–5[a]	1–5[a]	1–5[a]	1–5[a]	1–5[a]	1–5[a]	0 (0.00)	0 (0.0)
30–50	18–22[a]	42–46[a]	56–60[a]	62–66[a]	62–66[a]	61–65[a]	66–70[a]	71–75[a]	74 (26.33)	65 (25.3)
51–64	55 (46.6)	77 (43.3)	100 (44.1)	112 (42.7)	119 (42.2)	122 (42.4)	127 (43.9)	124 (41.9)	122 (43.4)	117 (45.5)
65+	40 (33.9)	54 (30.3)	66 (29.1)	83 (31.7)	96 (34.0)	100 (34.7)	91 (31.5)	96 (32.4)	85 (30.3)	75 (29.2)
Sex, n (%)										
Female	94 (79.7)	146 (82.0)	183 (80.6)	211 (80.5)	221 (78.4)	233 (80.9)	237 (82.0)	240 (81.1)	224 (79.7)	206 (80.2)
Male	24 (20.3)	32 (18.0)	44 (19.4)	51 (19.5)	61 (21.6)	55 (19.1)	52 (18.0)	56 (18.9)	57 (20.3)	51 (19.8)

[a] Exact counts suppressed for privacy reasons. ILD: interstitial lung disease; SSc: systemic sclerosis.

Table 2. Prevalent SSc and SSc-ILD cases.

Fiscal Year	Prevalent SSc Cases by Diagnosis Year, n	Cumulative Prevalent Cases of SSc[a], n	Prevalent SSc-ILD Cases by Diagnosis Year, n	Cumulative Prevalent Cases of SSc-ILD[a], n
2008/9	497	497	118	118
2009/10	381	823	75	178
2010/11	334	1073	70	227
2011/12	281	1239	62	262
2012/13	290	1435	50	282
2013/14	267	1590	44	288
2014/15	271	1749	39	289
2015/16	272	1909	43	296
2016/17	257	2007	38	281
2017/18	261	2114	20	257

[a] Cumulative prevalence is the sum of patients that are alive with the indication at the start of that year who are recorded that year and were diagnosed in any of the eligible cohort years. For example, cumulative prevalent cases in 2010 refers to all patients alive on April 1, 2010, diagnosed between 2008 and 2010. ILD: interstitial lung disease; SSc: systemic sclerosis.

Table 3. Overall prevalence of patients with SSc and SSc-ILD per 100,000 persons.

Fiscal Year 2017/18	SSc, n = 2114	SSc-ILD, n = 257	SSc Cases With ILD, %
Overall, n	19.1	2.3	12.0
Age group, yrs, n			
18–29	3.6	0.0	–
30–50	14.1	1.7	12.1
51–64	28.8	4.2	14.6
65+	29.4	3.2	10.9
Sex, n			
Female	31.2	3.6	11.5
Male	6.2	1.0	16.1

ILD: interstitial lung disease; SSc: systemic sclerosis.

per 100,000 persons) compared with males (6.2 per 100,000 persons). Of the 257 SSc-ILD cases, cumulative prevalence at the start of fiscal 2017/18 was found to be 2.3 per 100,000 persons. Unlike the SSc cohort, patients with SSc-ILD between the ages of 51 and 64 had the highest cumulative prevalence (4.2 per 100,000 persons). Females had a higher prevalence rate at the start of fiscal year 2017/18 (3.6 per 100,000 persons) compared with males (1.0 per 100,000 persons).

For the sensitivity analysis, the cumulative prevalence at the start of fiscal year 2017/18 was determined to be 2.3 cases per 100,000 persons. Patients aged 65+ had the highest cumulative prevalence (3.2 per 100,000 persons), and females had a higher prevalence of SSc-ILD at the start of fiscal 2017 (3.6 per 100,000 persons). Sensitivity analysis results can be found in Supplementary Table 2 (available with the online version of this article).

Survival results. Survival was calculated based on all identified cases during the 10-year study period. For patients with SSc, there were 1150 deaths (37.0%) in the population over the duration of the 10-year study period. The survival rates at 1, 5, and 10 years after diagnosis were 85.0%, 64.5%, and 44.9%, respectively. Figure 2 presents the Kaplan-Meier survival curve for patients with SSc, along with the proportion of patients at risk, those who died, and those who were censored.

Patients with SSc-ILD had lower survival rates and higher proportions of risk. There were 336 patients (63.7%) who died over the study period. The survival rates at 1, 5, and 10 years after diagnosis were 77.1%, 44.4%, and 22.0%, respectively. Figure 3 presents the Kaplan-Meier survival curve for patients with SSc-ILD, along with the proportion of patients at risk, those who died, and those who were censored.

DISCUSSION

This study determines the SSc and SSc-ILD prevalence and mortality in the total population in Ontario, Canada, which has universal healthcare access.

There is a paucity of information on prevalence estimates for SSc and SSc-ILD that have been published globally. A systematic review conducted by Bergamasco, *et al* examined the epidemiology of SSc and SSc-ILD including prevalence and survival from a number of studies in Europe and in North America.[16] They examined 39 studies from Europe and North America and found a wide variation in the reported prevalence estimates for SSc (7.2–33.9 per 100,000 persons and 13.5–44.3 per 100,000 persons, respectively). The investigators were able to derive prevalence estimates for SSc-ILD from only 1 study from Norway, where the estimated prevalence was found to be in the range of 1.7–4.2 per 100,000 persons. We found that the prevalence of SSc-ILD estimates from our study fell within this range (2.3 per 100,000 persons). The investigators also examined multiple sites in the US and Canada; however, North American SSc-ILD prevalence results were not available.[16]

There are some Canadian studies that have previously reported on the prevalence of SSc and SSc-ILD. One of the first Canadian studies that explored prevalence in a small setting by Thompson, *et al* estimated prevalence of SSc to be 28 cases
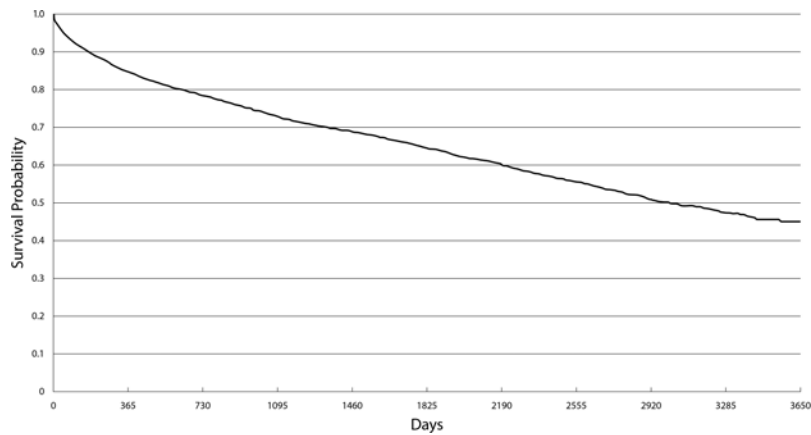
Figure 2. Kaplan-Meier survival curve and risk tables for patients with systemic sclerosis.
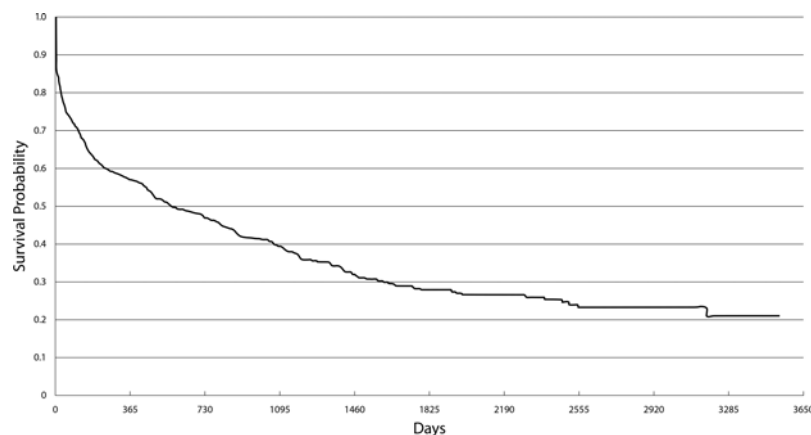

Figure 3. Kaplan-Meier survival curve and risk tables for patients with systemic sclerosis with interstitial lung disease.

per 100,000 persons.[6] However, the authors acknowledge that the results are not truly reflective of the larger population, as results from 2 of the 3 communities that they examined did not reach statistical significance. Bernatsky, *et al* analyzed data (1989–2003) from Quebec to estimate the prevalence of SSc using physician billing and hospitalization information.[24] They found the overall prevalence in 2003 to be 44.3 per 100,000 persons, also higher than our estimates. Similarly, a study from Alberta found that the SSc prevalence in 2007, using physician billing records and hospitalization data, was 57.7 per 100,000 females, and 9.8 cases per 100,000 males.[25] Another study from Canada reported that the sex ratio of patients with SSc was 4.7 females to 1 male, where males had more diffuse cutaneous SSc subset, more ILD, and reduced survival rates.[26] These findings are similar to those of our current study with respect to the sex distribution of cases and reduced survival in SSc-ILD. Our study used data from outpatient hospital clinics and inpatients, and considered only ICD-10-CA diagnosis codes. It is likely that higher prevalence estimates determined in other Canadian studies captured more community cases of SSc in physician billing information.

In our study, the probability of survival for patients with SSc was found to be lower/shorter than what has been previously reported. The systematic review conducted by Bergamasco, *et al*[16] found that in Europe, the survival rate of patients with SSc at 5 and 10 years was between 83–84% and 65–73%, respectively, whereas in North America, 10-year survival ranged between 66–82%. A second systematic review and metaanalysis, including 43 studies over the years 1964 to 2005 conducted in 12 countries, suggests 5- and 10-year SSc survival rates are 75% and 63%, respectively.[27] In contrast, our study found lower survival estimates, with 5- and 10-year survival rates in SSc at 65% and 45%, respectively. Given that our patient population was selected from inpatients and hospital-based outpatient clinics, we believe that patients might have had more severe SSc than patients in the studies that were included in the other systematic reviews and metaanalyses.

Our study found that approximately 12.0% of patients diagnosed with SSc were further formally diagnosed as having SSc-ILD, and that these patients had lower survival rates than overall SSc cases. When compared to survival data from a single tertiary care center in the US that reported 100% survival at 1 year and 77% at 5 years, where only 24 patients had SSc-ILD,[28] our results showed lower survival. To put our SSc findings into context, SSc-ILD survival rates are in line with those of multiple myeloma, a well-known cancer that has a similar 5-year survival rate (44%).[29]

Distribution by sex in our study was found to be in line with the expected distribution of approximately 80% women to 20% men affected by SSc and SSc-ILD, and aligned with reported distributions in other published studies.[7,8]

There are some notable strengths in this study. Our dataset is large and representative, as the denominator included the entire adult population of Ontario. Using administrative data from Ontario allows for population-level data to be examined without imputation and without extrapolation of results. SSc and SSc-ILD definitions derived using ICD-10-CA codes were validated by clinicians across both rheumatology and respirology who have extensive experience in treating and diagnosing SSc-ILD. Ten years of population data were included in order to ascertain stability of estimates. A sensitivity analysis was performed for defining SSc-ILD.

There are some limitations to the study. First, the databases capture diagnoses that are ambulatory and hospital-based and may not reflect community cases. They also provide limited clinical outcomes; as the studies conducted in other provinces have shown, the inclusion of billing and claims data may capture a wider population of patients.[24,25] Despite the assumptions that were made to identify the patients with SSc and those with ILD, the prevalence and proportion with ILD align with other studies, adding face validity to the methods used in this study.

Our study was conducted to provide estimates of the prevalence and survival rates of SSc and SSc-ILD in order to provide updated evidence to support the burden of SSc-ILD for clinicians and for policymakers in health-system planning, as well as payers to support the assessment of SSc-ILD treatments.

This analysis adds valuable information about prevalence estimates and survival in patients with SSc and SSc-ILD in Canada's largest province, and to our knowledge, provides the first population-based estimates using administrative data of SSc-ILD in Canada. Our study suggests that patients with SSc-ILD have a lower survival rate than those with only SSc, and that survival continues to be poor for patients with SSc-ILD.

ACKNOWLEDGMENT

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