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The prevalence of systemic autoimmune rheumatic diseases in Canadian pediatric populations: administrative database estimates

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Abstract To estimate systemic autoimmune rheumatic disease (SARD) prevalence using administrative data for pediatric populations in four Canadian provinces. Physician billing claims and inpatient hospitalizations from Alberta, Manitoba, Quebec, and Saskatchewan were used to define cases aged ≤18 years with a SARD diagnosis code in: one or more hospitalization, two or more physician visits within 2 years and at least 2 months apart, or one or more physician visit to a rheumatologist. Estimates ranged from 15.9/100,000 in Quebec [95 % confidence interval (95 % CI) 14.1, 18.0] to 23.0/100,000 in Manitoba (95 % CI 17.9, 29.2). SARDs were more common in females than in males across all provinces. There was a slightly higher prevalence among those living in urban compared to rural areas of Alberta (rate difference 14.4, 95 % CI 8.6, 20.1) and Saskatchewan (rate difference 13.8, 95 % CI 1.0, 26.6). Our results provide population-based prevalence estimates of pediatric SARDs in four Canadian provinces.

Keywords Pediatric rheumatic diseases · Systemic autoimmune rheumatic diseases · Epidemiology · Disease prevalence

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Introduction

Population-based administrative health databases are being increasingly used for outcomes research and surveillance in chronic rheumatic disease [1–4], but most studies have focused on adult populations. We used provincial administrative data to estimate systemic autoimmune rheumatic disease (SARD) prevalence in the pediatric population of four Canadian provinces: Alberta, Manitoba, Quebec, and Saskatchewan.

Methods

This study used physician billing claims and hospitalization records to ascertain SARDs cases. Physician services are recorded provincially for remuneration purposes and contain a single diagnosis, while hospitalization records contain up to 25 diagnoses, all of which were reviewed for SARDs (Table 1). Diagnoses are coded according to the World Health Organization's International Classification of Diseases (ICD), version 9, ICD 9-CM, or ICD 10 Canadian version (10-CA).

Data from the provinces spanned the following periods: Alberta 1994–2007, Manitoba 1995–2009, Quebec 1994–2003, and Saskatchewan 1998–2007. We used a case ascertainment algorithm previously validated in adult SARDs [5], which defines cases as one or more of: (a) $\geq$1 hospitalization with a SARD diagnosis recorded anywhere on the hospital discharge record, (b) $\geq$2 physician visits with a SARD diagnosis at least 2 months apart but within a 2-year period, (c) $\geq$1 physician visit to a rheumatologist with a SARD diagnosis. A subject was included in our estimates if he/she remained in the province and was $\leq$18 years of age as of the end of the study interval. Cases that satisfied more than one definition were only counted once in prevalence estimates.

Statistics Canada Census data from the last year of the study period for each province were used to estimate prevalence per 100,000 residents aged $\leq$18 years. Urban regions were defined as census metropolitan (population >100,000) and census agglomeration (population >10,000 but <100,000) areas and were determined by residence code in Saskatchewan and postal code in Alberta, Quebec, and Manitoba. All other areas were considered rural. Residence codes in Saskatchewan are five digit numeric codes that uniquely identify each city, town, village, rural municipality, and First Nation. We did not provide estimates of individual SARDs (systemic lupus, scleroderma, or inflammatory myopathies), because we were not able to distinguish these by diagnosis code for Manitoba and Saskatchewan.

Results

Table 2 presents the crude SARDs prevalence estimates per 100,000 individuals $\leq$18 years. The overall pediatric SARDs prevalence estimates per 100,000 were 19.6 in Alberta [95 % confidence interval (95 % CI) 16.7, 22.9], 23.0 in Manitoba (95 % CI 17.9, 29.2), 15.9 in Quebec (95 % CI 14.1, 18.0), and 20.9 in Saskatchewan (95 % CI 15.6, 27.5). The 95 % CI of the estimated differences in rates between Manitoba and Saskatchewan, Manitoba and Alberta, Saskatchewan and Alberta, and Saskatchewan and Quebec were too wide for definitive conclusions; however, there was evidence for a difference in rates between Quebec versus Manitoba (-7.1 per 100,000, 95 % CI -12.9, -1.3), and Quebec versus Alberta (-3.7, 95 % CI -7.2, -0.1). SARDs were more common in females than in males across all provinces. There was a slightly higher prevalence among those living in urban compared to rural areas of Alberta (rate difference 14.4, 95 % CI 8.6, 20.1) and Saskatchewan (rate difference 13.8, 95 % CI 1.0, 26.6). The rate difference intervals for urban compared to rural areas of residence in Manitoba and Quebec had CIs that were too wide for definitive conclusions.

Discussion

We calculated population-based estimates of pediatric SARDs prevalence in four Canadian provinces, with estimates suggesting about 2 SARDs cases per 10,000 residents aged 18 or less. We did not provide estimates of individual SARDs because we were not able to distinguish these by diagnosis code for Manitoba and Saskatchewan. Pediatric SARDs prevalence estimates from our study are higher than those reported by the only pediatric rheumatology clinic in Saskatchewan, Canada [6], and prevalence rates have not been previously reported for Alberta, Manitoba, or Quebec. Our case definition aims for adequate sensitivity, which may have resulted in lower specificity than clinic-based data, and consequently misclassification of SARDs cases leading to a higher prevalence rate. For example, during the time period of this study in Saskatchewan, pediatric rheumatology billed as general pediatrics, and two general pediatricians could not bill for the same diagnosis code for the same patient on the same day, which may have contributed to misclassification of administrative

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Table 1 ICD 9 and corresponding ICD 10-CA codes included as SARDs

ICD 9 code	Diagnoses	ICD 10-CA code	Diagnoses
710.0	Systemic lupus erythematosus Disseminated lupus erythematosus Libman–Sacks disease	M32.1	Systemic lupus erythematosus, with organ or system involvement
		M32.8	Other forms of systemic lupus erythematosus
		M32.9	Systemic lupus erythematosus, unspecified
710.1	Systemic sclerosis Acrosclerosis CRST syndrome Progressive systemic sclerosis Scleroderma	M34.x	Systemic sclerosis (scleroderma)
710.2	Sicca syndrome Keratoconjunctivitis sicca Sjögren's disease	M35.0	Sicca syndrome (Sjögren)
710.3	Dermatomyositis Poikilodermatomyositis	M33.x	Dermatopolymyositis
	Polymyositis with skin involvement	M36.0	Dermato(poly)myositis in neoplastic disease
710.4	Polymyositis	M33.x	Dermatopolymyositis
		M36.0	Dermato(poly)myositis in neoplastic disease
710.5	Eosinophilia myalgia syndrome Toxic oil syndrome	None	None
710.8	Other specified diffuse diseases of connective tissue Multifocal fibrosclerosis (idiopathic) NEC Systemic fibrosclerosing syndrome	M35.8	Other specified systemic involvement of connective tissue
710.9	Unspecified diffuse connective tissue disease Collagen disease NOS	M35.9	Systemic involvement of connective tissue, unspecified

710.6 and 710.7 are not used in ICD 9

data diagnoses. On the other hand, a clinic-based estimate could potentially underestimate prevalence (if SARD cases do not come to the attention of the pediatric rheumatology clinic).

In reports based on clinic data in BC, 3–6 % of pediatric rheumatology patients seen in follow-up are SARDs cases [7]. The most common pediatric SARD is systemic lupus erythematosus (SLE), which accounts for about a third of SARDs cases, while juvenile dermatomyositis accounts for approximately another fifth. SARD prevalence across regions may be driven in part by differences in race/ethnicity. A study from British Columbia, Canada [8] using clinical records suggested a prevalence of 3.3 cases per 100,000 in non-First Nations children, with a much higher rate in First Nations (Native North American) children of 8.8 cases per 100,000. SLE is more frequent (and diagnosed at a younger age) in non-white individuals, specifically in Hispanics, blacks (both African-Americans and Afro-Caribbeans), Asians, and persons of Native North American origin [9–11]. Within the provinces studied, Manitoba and Saskatchewan have more residents of Aboriginal status. Aboriginal status includes Native North American, Métis (part European and part Native North American), and Inuit (from Northern Canada). In Manitoba and Saskatchewan, 15.8–14.9 % of the population are Aboriginal, respectively, by self-report according to Statistics Canada, compared to 5.8 % in Alberta and 1.5 % in Quebec [12]. The higher prevalence estimates from Manitoba and Saskatchewan may be driven by the large number of Aboriginal residents in these provinces. Our study was not able to examine the relationship between Aboriginal status and pediatric SARDs prevalence rates directly because we did not have a reliable indicator of Aboriginal status in any province other than Alberta.

We observed higher rates of SARDs in pediatric residents of urban Alberta compared to rural areas in that province. Although data in Saskatchewan suggested a similar trend, due to the low numbers of male cases, the differences were not statistically significant for rural and urban regions when stratified by sex. Interestingly, at least one report has also suggested a higher occurrence also for juvenile idiopathic arthritis in urban populations [13]. The reasons for such a phenomenon are unknown. It is possible that an apparent higher disease prevalence in urban versus

Table 2 Estimated prevalence of systemic autoimmune diseases per 100,000 individuals ≤18 years of age using Canadian provincial administrative health data

Province	Overall (95 % CI)			Females (95 % CI)			Males (95 % CI)		
	Total	Rural	Urban	Total	Rural	Urban	Total	Rural	Urban
Alberta	19.6 (16.7, 22.9)	11.2 (7.9, 15.3)	25.6 (21.2, 30.5)	27.7 (22.8 33.4)	16.3 (10.8, 23.8)	35.8 (28.5, 44.3)	11.9 (8.8, 15.7)	6.3 (3.1, 11.3)	15.8 (11.3, 21.6)
Manitoba	23.0 (17.9, 29.2)	26.2 (18.3, 36.4)	20.4 (14.1, 28.7)	34.0 (25.2, 45.0)	40.0 (26.2 58.6)	29.1 (18.4, 43.7)	12.6 (7.6, 19.6)	13.1 (6.0, 24.9)	12.1 (5.8, 22.3)
Quebec	15.9 (14.1, 18.0)	15.5 (12.5, 18.9)	16.2 (13.9, 18.9)	23.4 (20.2, 27.0)	20.5 (15.6, 26.3)	25.1 (21.0, 29.8)	8.8 (6.9, 11.0)	10.7 (7.4, 15.0)	7.7 (5.5, 10.4)
Sask[a]	20.9 (15.6, 27.5)	15.7 (10.1, 23.4)	29.6 (19.5, 43.0)	32.8 (23.3, 44.8)	25.6 (15.4, 39.9)	44.9 (27.4, 69.3)	9.6 (5.0, 16.8)	6.4 (2.1, 14.9)	15.0 (6.0, 30.8)

[a] Saskatchewan

rural areas is at least partially linked to access to care, since cases must seek medical attention in order to be included in the prevalence estimates.

In general, SARDs are more common in females than in males. For example, in children, SLE has been reported to have a prevalence ratio of about four or five females to every male case [14–16], and in juvenile dermatomyositis at 2.2 to 1 [17]. The female to male ratios that we estimated are consistent with this phenomenon.

Compared to estimates in adults, prevalence estimates for pediatric rheumatic diseases are relatively rare. Accurate estimates of rates of pediatric-onset chronic disease are critical to determine the public health impact of these disorders. Much of the existing literature on outcomes in patients with rheumatic disease is based on data from tertiary care centers, which is limited by sample size and the potential for selection bias. An alternative approach is the creation of relatively large population-based cohorts from administrative databases.

Prevalence estimates were based on algorithms previously developed in the adult population [5, 18] that have not been validated in the pediatric setting. Recent guidelines [19] recommend age-specific validation studies to account for differences in the sensitivity and specificity of case finding algorithms across ages, as there is the potential for misclassification of cases, which may result in biased prevalence estimates. Additionally, some physicians may code cases for which they were asked to "rule out" a diagnosis as that diagnosis, which would also contribute to misclassification of SARDs cases, increasing prevalence estimates. It is not clear how shadow billing and the alternate funding plans (salary based) through which many Canadian pediatric rheumatologists are currently reimbursed would affect prevalence estimates, and we could not account for this. Additionally, in some provinces, pediatric rheumatologists bill as general pediatricians or internists, and we would not have captured these individuals as rheumatologists in our algorithm. A validation study was beyond the scope of this work. Our analysis also did not account for potential imperfect sensitivity or specificity of the information contained in the databases; latent class models have been used for SARD estimation in adults [4, 20], but their use has been limited for prevalence estimation in pediatric populations, since these methods are difficult to apply in settings where the absolute number of cases is relatively small (as is the case for pediatric SARDs).

Despite some limitations, our prevalence estimates provide population-based prevalence estimates of SARDs in the Canadian pediatric population. These data can provide useful information about potential regional and demographic variations in disease estimates. Pediatric validation studies will be important to assess the quality of these estimates and determine the optimal case definition(s) for future work.

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Conflict of interest None.

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