

Sickle Cell Disease in Ontario: An Epidemiologic Profile Based on Health Systems Administrative Data

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Geographical variables were adapted from Statistics Canada, PCCF+ 2006, PCCF+ 2011, PCCF+ 2016. This does not constitute an endorsement by Statistics Canada of this product.

All authors approved the final manuscript submitted and contributed to the conception and design of the study, interpretation of the data, and critical revisions of the manuscript for important intellectual content. JP drafted the initial manuscript and EK carried out the statistical analysis. No authors have any relevant conflicts of interest to declare.

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4 *Abstract*
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9 Background: The number of sickle cell patients in Ontario is unknown. In the absence of a
10 formal registry, it may be possible to determine an approximate census via analysis of health
11 services administrative databases.
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16 Methods: Ontario patients with a diagnosis of sickle cell disease were identified through
17 queries of the Discharge Abstract Database, the National Ambulatory Care Reporting System,
18 and Newborn Screening Ontario database. The period of inquiry was April 1st 2007 through
19 March 31st 2017. Repeat interactions by the same patient were identified by cross-referencing
20 unique, individual Ontario Health Insurance Plan numbers.
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28 Results: Health system interactions were documented for 3 418 individuals with sickle cell
29 disease, with 56% female, and a median age of 24 years. Over the 10-year period of study,
30 patients were hospitalized a median of 1 time (Interquartile Range [IQR] 1-5, range 0-148), and
31 visited the emergency department twice (IQR 1-7, range 0 – 1 125) for sickle cell disease. Of
32 the 229 (6.7%) of patients who died during the period of study, the average age at death was 55
33 years. Most patients lived in urban areas, with approximately 40% in low income
34 neighbourhoods. The annual mortality rate remained stable over the period of study while the
35 number of affected births declined.
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48 Interpretation: Patients with sickle cell disease have a population prevalence in Ontario of
49 approximately 1 in 4 200 and have a significant burden of disease. Similar queries of health
50 services administrative databases may be feasible in other Canadian provinces.
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Introduction

Sickle cell disease is an autosomal recessive condition and the most common monogenetic disease in the world, affecting approximately 25 million individuals and with 300-400 000 affected children born each year.¹ The most common genotypes are homozygous hemoglobin S, and compound heterozygosity of hemoglobin S with either β -thalassemia or hemoglobin C.² It is especially prevalent in low-resource nations where, due the protective effect of being a HgbS carrier, malaria is endemic.³ With increasing immigration, however, a growing number of individuals with sickle cell disease are now found in higher-resource countries of the Northern hemisphere.⁴ In these settings, the provision of simple interventions such as prophylactic antibiotics, hydroxyurea and blood transfusion have resulted in dramatic increases in life expectancy, which now approaches 60 years of age.⁵ However, affected individuals still suffer from significant morbidity due to intermittent vaso-occlusive and hemolytic crises, and progressive organ dysfunction.⁶ In the United States, the burden of sickle cell disease has been estimated to cost the health care system over US\$1 billion each year.⁷

Given the high degree of morbidity and associated health care costs associated with a diagnosis of sickle cell disease, an epidemiologic profile, including population prevalence estimates and associated demographics of cases, would be of value in resource allocation planning (i.e., investment in additional comprehensive care programs). At the time of submission, there were no registries of sickle cell patients in Canada with which to ascertain patient numbers and burden of disease. The frequency in which patients with sickle cell disease access acute and emergency care, however, provides an opportunity to enumerate the

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3 number of individual patients in a defined geographic area. The need for such information was
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5 highlighted by the Sickle Cell Awareness Group of Ontario (SCAGO), a charitable patient
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7 organization advocating for patients with sickle cell disease, who were engaged in all aspects of
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9 this project. The current study represents an attempt to approximate the number of sickle cell
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11 patients in Ontario, Canada's most populous province, by querying inpatient and outpatient
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13 administrative health databases over a 10-year period.
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21 *Methods*

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26 We used health services administrative databases to search for patients with a diagnosis of
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28 sickle cell disease in Ontario during the time period April 1st 2007 through March 31st 2017.
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30 Datasets were linked using unique encoded identifiers and analyzed at ICES. The first was the
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32 Discharge Abstract Database (DAD), compiled by the Canadian Institute for Health Information
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34 (CIHI), and containing clinical (diagnoses and procedures/interventions), demographic, and
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36 administrative information for all admissions to acute care hospitals in Ontario. Secondly, the
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38 National Ambulatory Care Reporting System (NACRS), also compiled by CIHI, was searched for
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40 similar data elements for all patient visits made to hospital- and community-based ambulatory
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42 care centres (emergency departments (ED), cancer care clinics, and hemodialysis units) in
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44 Ontario; and used to compile a database of all visits to Same-Day Surgery (SDS) clinics. These
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46 databases were queried for ICD-10 codes indicating a diagnosis of sickle cell disease.
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49 To supplement these records, a third data source, Newborn Screening Ontario, was also
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51 queried for diagnoses of sickle cell disease made shortly after birth. To identify repeat health
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3 care interactions for sickle cell disease by the same patient, individuals were cross-referenced
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5 by unique, encoded Ontario Health Insurance Plan (OHIP) number. This identifier in turn
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7 allowed a query of the Registered Persons Database (RPDB), which includes basic demographic
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9 information (age, sex, postal code, date of birth, and date of death for deceased individuals) for
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11 all Ontario residents covered by OHIP (i.e., the publicly funded provincial health system). See
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13 supplemental appendix for additional details.
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18 The assembled cohort of patients with sickle cell disease were then analyzed
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20 descriptively regarding age, sex, location of residence (urban vs rural residence and regional
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22 healthcare zone), and both type and number of health care interactions for sickle cell in our 10-
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24 year study period. The rate of natural increase was calculated each year by subtracting the
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26 number of deaths from the number of newborn diagnoses. The number sickle cell disease-
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28 related acute and emergency health care interactions by those without Ontario Health
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30 Insurance Plan (OHIP) coverage (i.e., individuals who are neither Canadian citizens, permanent
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32 residents, nor landed immigrants or refugees)⁸ were assessed but otherwise not included in the
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34 calculations of disease prevalence due to the inability to distinguish individual patients who
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36 lacked an OHIP number to use as a unique identifier. To determine the robustness of this
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38 analysis, cohort size was recalculated after decreasing the period of analysis from 10 to 5 and
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40 then 2 years, and with the sequential exclusion of data from NSO (newborn screening) and
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42 NACRS (outpatient interactions).
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49 The use of the data in this project is authorized under section 45 of Ontario's Personal
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51 Health Information Protection Act and does not require review by a Research Ethics board.
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Results

A total of 44 770 patient records with a diagnosis of sickle cell disease were identified between April 1 2007 through March 31st 2017; of these, 1 168 (2.6%) did not have an associated OHIP number. While the number of individuals represented by these non-OHIP visits could not be determined due to lack of a unique identifier, their occurrence declined over time (see Figure 1). Non-OHIP interactions were proportionally highest in the Central Toronto and Hamilton regions (8.5% and 7.5% of interactions respectively, data not shown).

The remaining 43 602 healthcare interactions were cross-referenced by OHIP number, revealing a total of 3 418 unique individuals with a median age of 24 years (IQR 9-39) at time of first health care interaction for sickle cell disease during the period of study. Most patients were female (54%), although males predominated in the age cohort younger than 15 years (Figure 2). During the 10 years of the study period 229 (6.7%) of these individuals were documented to have died, with mean age of death 55 years (SD 21 years, range 2 to 93). Average age at time of death remained stable over the 10-year period of study. Also during this time period, 492 newborns were identified, representing an average rate of natural increase of 35 individuals per year (Figure 3). Of these, 292 (59%) were Hemoglobin (Hgb)-SS, 130 (26%) were Hgb-SC, 53 (1.6%) were Hgb-S β , 7 (0.2%) were Hgb-SE and 7 (0.2%) were Hgb-S/HPFH.

While sickle cell patients were found throughout the province, they were heavily concentrated in urban areas of Toronto, Hamilton and Ottawa (Figure 4), and almost double the anticipated proportion of the cohort lived in the lowest income quintile, as compared to the

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3 general Canadian population. While 25% of families with affected newborns changed postal
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5 codes within one year of birth, none moved from a rural to an urban area.
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8 The pattern of healthcare utilization by this population was highly variable (Table 1).
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10 Over the 10-year period of review, a total of 14 985 inpatient admissions and 22 800 ED
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12 department visits for sickle cell disease were documented; an additional 4 957 non-ER
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14 outpatient visits were also documented. The median number of ED visits per patient was 2 (IQR
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16 1-7) but was heavily skewed by a small number of high-utilization individuals. Similarly, while
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18 the median number of hospitalizations over the 10 period was only 1 per patient (IQR 1-5) the
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20 range was very large (0 to 148). Slightly more male than female patients were identified by
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22 newborn screening (52% vs 48%), and male patients had slightly higher rates than females of
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24 both ED visits and inpatient admissions.
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30 Sensitivity analysis revealed that while excluding analysis of non-ED outpatient visits had
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32 little effect on the total number of patients captured, limiting analysis to inpatient visits
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34 decreased the number of identified patients by nearly one quarter. There was a similarly large
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36 effect when the period of analysis was reduced from 10 to 5, and from 5 to 2 years (Figure 5)
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42 *Interpretation*

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47 Ontario has a population of 14 million people, is ethnically diverse, and virtually all individuals
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49 in Ontario have universal access to hospital care and physician services through the publicly
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51 funded provincial health system (e.g., OHIP). The estimated census of 3 418 individuals with
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53 sickle cell disease living in Ontario during this period of study suggests a population prevalence
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3 of 1 in 4 200 Ontarians. While this meets Health Canada's definition of a rare disease (ie., fewer
4 than 5 in 10 000⁸), it is notable that sickle cell disease is in fact twice as prevalent as current
5 population estimates for other, better funded genetic conditions such as hemophilia⁹ and cystic
6 fibrosis.¹⁰ While there was a large degree of heterogeneity in the intensity with which patients
7 required acute care management, the absolute number of emergency department and
8 inpatient visits for sickle cell disease required by this cohort over the 10-year period of review
9 was high. The slight overrepresentation of male vs female patients in rates of birth, ED visits
10 and inpatient admissions is unexplained, although sex differences in the prevalence and
11 severity of autosomal alleles has also been documented in other conditions.¹¹
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25 The economic impact of sickle cell disease is considerable. One study from the United
26 States, for example, estimated the lifetime costs of a single patient with sickle cell disease to
27 approach US\$9 million.¹² While the equivalent costs in the context of the Canadian healthcare
28 system have yet to be assessed, they will likely increase over time. For example, while the
29 number of affected births in the current study appeared to be trending downwards over the
30 period of analysis, the rate of natural increase remained positive at approximately 35 new cases
31 per year, meaning that the number of affected individuals is expected to continue increasing.
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42 Amongst newborns, the incidence of Hgb-SC (generally considered a less severe form of sickle
43 cell disease) was approximately half that observed for Hgb-SS, consistent with what has been
44 reported in the United States.² In addition, although the average age at death in this cohort
45 was 55 years, also comparable to what has been estimated in the United States,¹³ the average
46 patient age was only 24, suggesting that the burden of disease in this population will increase
47 over time as these patients age and accumulate progressive chronic organ dysfunction and
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3 need for transfusion support. While transfusion data was not captured in the current study,
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5 similar investigations in the United States have revealed a doubling in hospital stays between
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7 2000 and 2013 in which transfusion was the sole procedure,¹⁴ with one third admissions for
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9 pain crises now accompanied by transfusion.¹⁵ This growing reliance on transfusion support of
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11 patients with sickle cell disease may reflect an increasing incidence of disease complications for
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13 which transfusion as indicated, but may also reflect increased awareness of transfusion
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15 guidelines.^{16,17,18}
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20 The analysis of health system administrative databases is a well-established
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22 methodologic technique for approximating the number of patients in a defined geographic area
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24 with sickle cell disease.¹⁹ An analysis of Medicaid claims, for example, has been used to
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26 enumerate the number of pediatric sickle cell patients in Tennessee, Michigan and Georgia.
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28 These studies have been validated using data from newborn screening databases and generally
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30 show a high sensitivity and specificity, although accuracy varied with the number of interactions
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32 required to confirm a case, and with the length of time the audit was conducted over.¹⁹ The
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34 current study did not require multiple interactions to confirm coding accuracy and therefore
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36 may be susceptible to false positives (eg., miscoding a patient with sickle cell trait as disease).
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38 However, important advantages of our study include capture of all insured patients within a
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40 single-payer, public health-care system; the inclusion of data from a universal newborn screen
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42 program to capture patients too young to have required treatment in an acute care facility; and
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44 a 10-year period of surveillance, allowing the capture of patients who infrequently require
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46 acute care. While this study did not capture patients without OHIP coverage, the small number
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48 of such interactions suggests that this was a small population. Patients who never sought acute
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3 care over the 10-year period of study would also not have been captured, but this, too, is likely
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5 a small number of patients. In one study, for example, only 12% of pediatric sickle cell patients
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7 never sought treatment over a 5-year period of follow-up.²⁰ It is also possible that patients
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9 lacking OHIP coverage are less likely to seek acute care for socioeconomic reasons, and are
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11 therefore present in larger numbers than the captured interactions with the healthcare system
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13 would suggest. Among patients who do have healthcare insurance, however, there is no
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15 evidence that lower socioeconomic status results in under-utilization of healthcare services. On
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17 the contrary, such patients tend to have greater reliance on acute care support due to
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19 inadequate access to preventative health care and increased exposure to environmental
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21 exacerbators of their disease.^{21,22}
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28 The methodology utilized in this study could be applied to other provinces in Canada,
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30 but with some important caveats. First, newborn screening must be in place to identify
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32 patients too young to have required inpatient or outpatient medical care for their diagnosis. At
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34 time of publication, newborn screening for sickle cell disease is performed in British Columbia
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36 (including Yukon Territory), Alberta (with Northwest Territories), Ontario (including Nunavut),
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38 Quebec, Nova Scotia, Prince Edward Island and New Brunswick, but the number of years of
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40 data available for analysis varies; Alberta, the most recent addition, only began newborn
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42 screening in 2019.²³ A more important consideration is the inclusion of data from outpatient
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44 visits. In Canada, this information is captured by the National Ambulatory Care Reporting
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46 System, but the degree to which this includes visits to emergency departments varies by
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48 province.²⁴ In addition, diagnosis and procedure codes are submitted with varying degrees of
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50 detail. While inpatient data submitted to the DAD is more consistently collected on a national
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3 level, results from this study suggest it will miss approximately one quarter of sickle cell
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5 patients when using a 10-year case ascertainment period. Shortening the period of analysis to
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7 less than 10 years would result in further loss of sensitivity. The duration of analysis required to
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9 sustain data accuracy may increase over time, as the continued provision of comprehensive
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11 care to sickle patients steadily decreases the frequency of emergency department visits and
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13 hospitalizations.²⁵
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18 A number of important limitations in this study are acknowledged, including the
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20 exclusion both of individuals lacking provincial health care insurance, and those with health
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22 coverage who had sufficiently mild disease that they required neither emergency or inpatient
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24 care during the 10-year period of analysis. In addition, the diagnostic codes used in the data we
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26 gathered have not yet been validated against individual patient chart review.
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32 *Conclusion*

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37 The number of individuals with sickle cell disease in Ontario is substantial and continues to
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39 increase. This study represents the first attempt to systematically enumerate the number of
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41 individuals in Canada with sickle cell disease, and in the absence of a formal patient registry, the
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43 methodology adapted likely represents the most accurate means of monitoring the prevalence
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45 of this disease. Estimates of sickle cell disease prevalence and burden can be used by advocacy
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47 groups, such as SCAGO, and government agencies to inform funding decisions, best care
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49 practices, and policy development.
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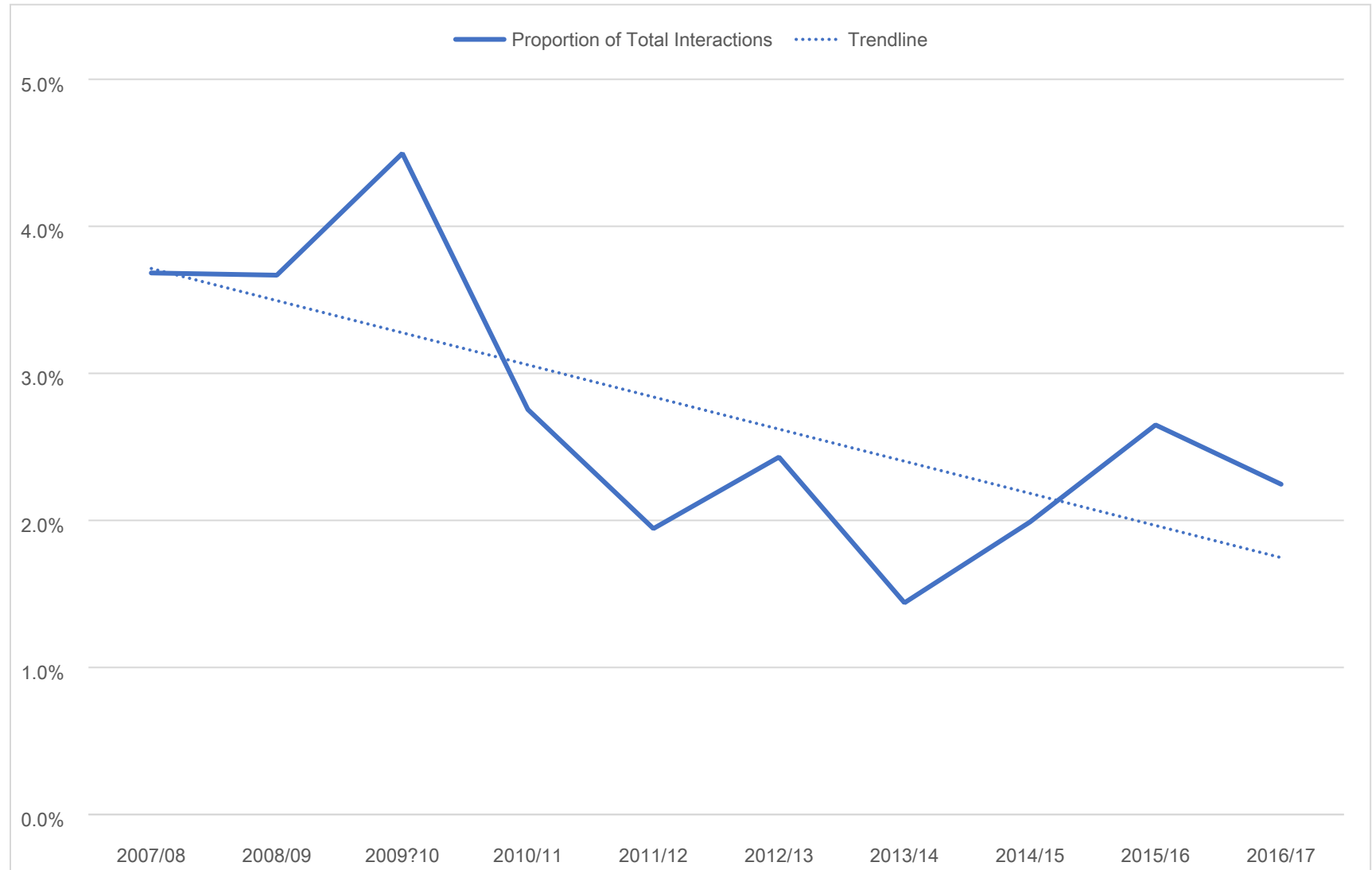
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Figure 1: Proportion of healthcare interactions for sickle cell disease by Individuals without insurance through the Ontario Health Insurance Plan (OHIP). Dashed line = linearized trendline



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Figure 2: Age Distribution of 3 418 Sickle Cell Patients Identified in Ontario, FY2007/08-2016/17

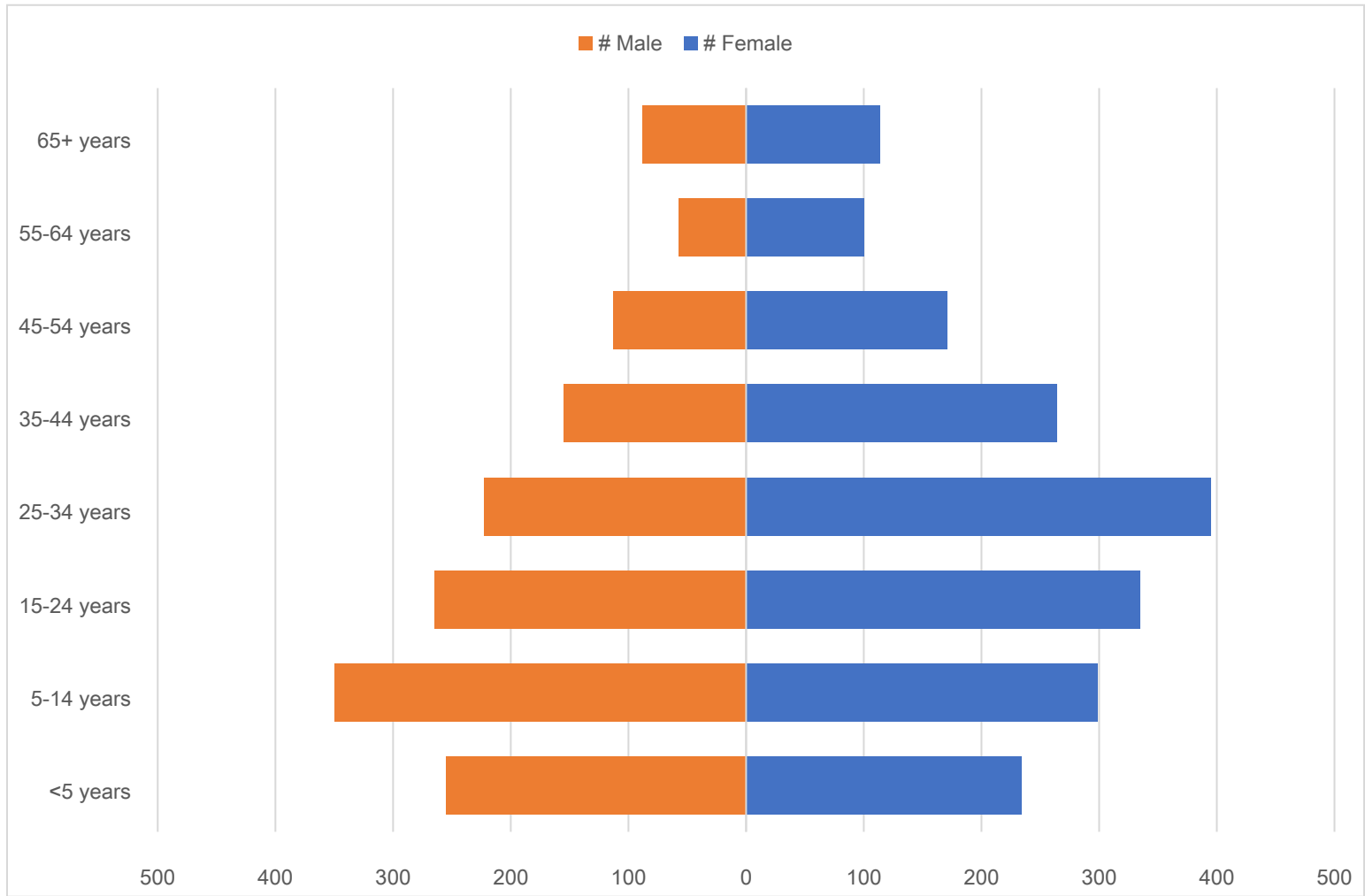
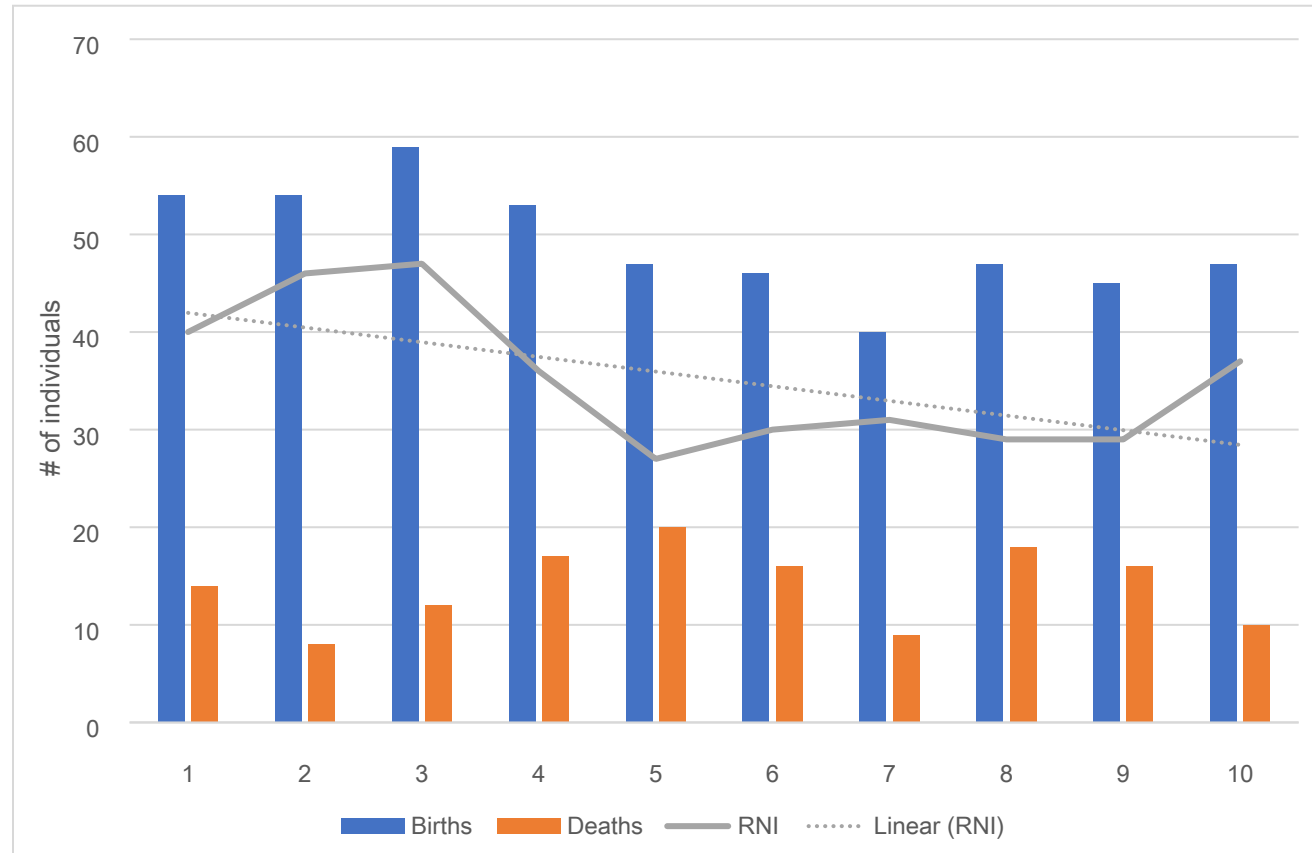


Figure 3: Estimated Population Change in Ontario Patients with Sickle Cell Disease Over Time



RNI = rate of natural increase

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Figure 4: Distribution by Geographic Area of Ontario Patients with Sickle Cell Disease

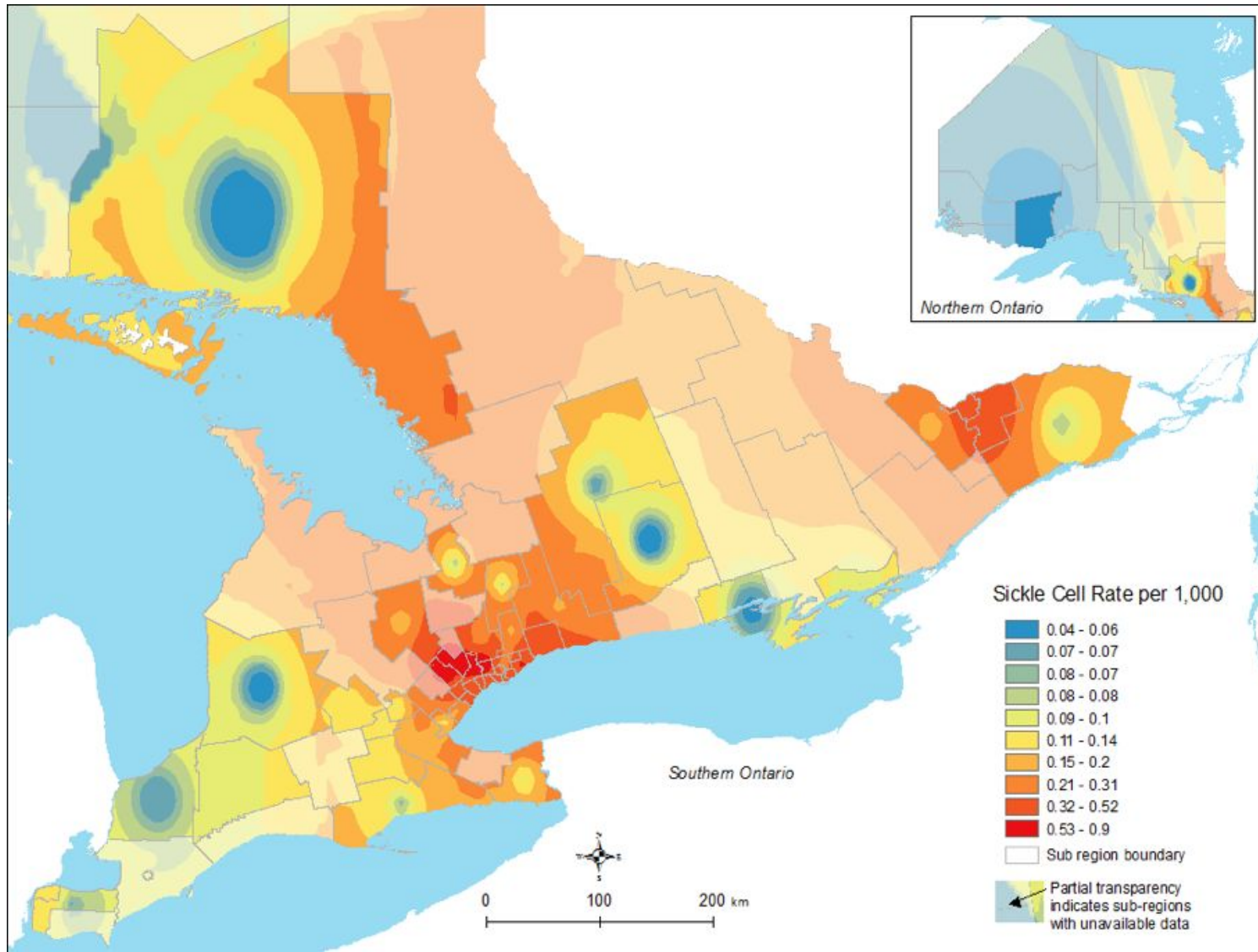


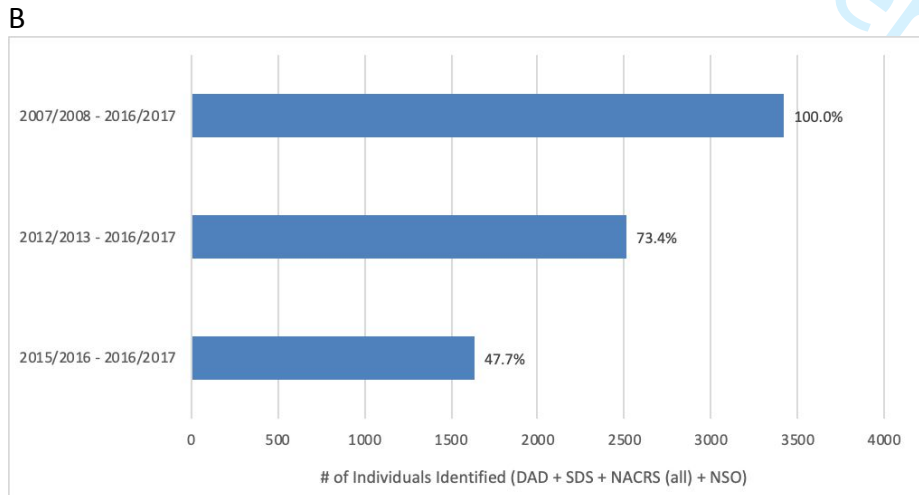
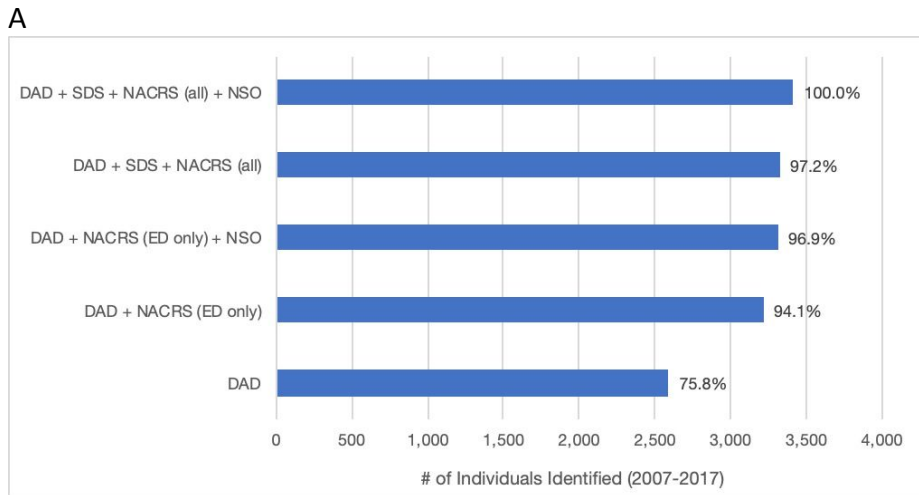
Table 1: Health care utilization for sickle cell disease among persons with any health care contact with a recorded diagnosis of sickle cell disease in Ontario, from FY 2007/08 to 2016/17

Type of health care contact		Female (N=1 912)	Male (N=1 506)	Total (N=3 418)
<i>Inpatient admissions (DAD)</i>	n (%)	1,443 (75.5%)	1,149 (76.3%)	2,592 (75.8%)
	Mean ± SD	4.05 ± 7.54	4.81 ± 9.64	4.38 ± 8.53
	Median (IQR)	1 (1-4)	2 (1-6)	1 (1-5)
	Range (min-max)	0-85	0-148	0-148
<i>Same day surgeries (SDS)</i>	n (%)	123 (6.4%)	89 (5.9%)	212 (6.2%)
	Mean ± SD	0.07 ± 0.30	0.09 ± 0.76	0.08 ± 0.55
	Median (IQR)	0 (0-0)	0 (0-0)	0 (0-0)
	Range (min-max)	0-4	0-26	0-26
<i>ED visits (NACRS)</i>	n (%)	1,366 (71.4%)	1,206 (80.1%)	2,572 (75.2%)
	Mean ± SD	5.45 ± 11.80	8.27 ± 37.92	6.69 ± 26.71
	Median (IQR)	1 (0-6)	2 (1-8)	2 (1-7)
	Range (min-max)	0-183	0-1,125	0-1,125
<i>Non-ED outpatient visits (NACRS)</i>	n (%)	121 (6.3%)	87 (5.8%)	208 (6.1%)
	Mean ± SD	1.32 ± 23.36	1.62 ± 31.36	1.45 ± 27.17
	Median (IQR)	0 (0-0)	0 (0-0)	0 (0-0)
	Range (min-max)	0-777	0-919	0-919
<i>Newborn screening (NSO)</i>	n (%)	238 (12.4%)	254 (16.9%)	492 (14.4%)

DAD = Discharge Abstract Database; NACRS = National Ambulatory Care Reporting System; SDS = Same Day Surgery; ED = Emergency Department; NSO = Newborn Screening Ontario

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Figure 5: Sensitivity Analysis by A) Health System Administrative Database Source and B) Years of Health System Administrative Database Analyzed



DAD = Discharge Abstract Database; NACRS = National Ambulatory Care Reporting System; SDS = Same Day Surgery; ED = Emergency Department; NSO = Newborn Screening Ontario

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6 *Supplement: Detailed methodology*
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10 Consecutive DAD records were linked together to form 'episodes of care' among the
11 hospitals to which patients have been transferred after their initial admission. Similarly,
12 NACRS records were linked with other data sources to identify transitions to other care
13 settings, Same Day Surgery clinics (SDS), inpatient acute care or psychiatric care. DAD,
14 NACRS, and SDS were queried for the following International Classification of Diseases, 10th
15 revision, Canadian Edition (ICD-10-CA) codes:
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- 18 • D570: Sickle cell anemia with crisis
 - 19 • D571: Sickle cell anemia without crisis
 - 20 • D572: Double heterozygous sickling disorders
 - 21 • D578: Other sickle cell disorders
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38 Newborn Screening Ontario was queried separately with the goal of identifying patients
39 diagnosed with sickle cell disease at birth. The NSO database is a publicly-funded program
40 NSO that screens almost every newborn in Ontario for rare but treatable diseases using a
41 combination of advanced laboratory techniques, and which expanded to include sickle cell
42 disease and traits in 2006. Newborns diagnosed with Hemoglobin SS, S β , SC, SE and
43 S/HPFH were identified through this database search.
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53 Repeat presentations by the same patient within each of the above databases were identified
54 by cross-referencing individual Ontario Health Insurance Plan (OHIP) numbers. Basic
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3 demographic details for each individual patient were available through their OHIP file,
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5 including their 6-digit post code on July 1st of each year of query. This postal code was in
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8 turn linked to the Postal Code Conversion File (PCCF+ 2006, 2011 and 2016) to obtain
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10 additional census geographic identifiers such as urban/rural residence flag (living in a
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12 community with less than 10 000 individuals) and neighbourhood income quintile.
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