Validation of algorithms to determine incidence of Hirschsprung disease in Ontario, Canada: a population-based study using health administrative data

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Objective: Incidence rates of Hirschsprung disease (HD) vary by geographical region, yet no recent population-based estimate exists for Canada. The objective of our study was to validate and use health administrative data from Ontario, Canada to describe trends in incidence of HD between 1991 and 2013.

Study design: To identify children with HD we tested algorithms consisting of a combination of diagnostic, procedural, and intervention codes against the reference standard of abstracted clinical charts from a tertiary pediatric hospital. The algorithm with the highest positive predictive value (PPV) that could maintain high sensitivity was applied to health administrative data from April 31, 1991 to March 31, 2014 (fiscal years 1991–2013) to determine annual incidence. Temporal trends were evaluated using Poisson regression, controlling for sex as a covariate.

Results: The selected algorithm was highly sensitive (93.5%) and specific (>99.9%) with excellent predictive abilities (PPV 89.6% and negative predictive value >99.9%). Using the algorithm, a total of 679 patients diagnosed with HD were identified in Ontario between 1991 and 2013. The overall incidence during this time was 2.05 per 10,000 live births (or 1 in 4,868 live births). The incidence did not change significantly over time (odds ratio 0.998, 95% confidence interval 0.983–1.013, p = 0.80).

Conclusion: Ontario health administrative data can be used to accurately identify cases of HD and describe trends in incidence. There has not been a significant change in HD incidence over time in Ontario between 1991 and 2013.

Keywords: Hirschsprung disease, algorithm validation, incidence, health administrative data

Introduction

HD is a congenital disease in which a section of the bowel is aganglionic, beginning at the internal anal sphincter and extending proximally for varying lengths through the colon.1 Due to the impaired physiology of the nerves in this area, the affected segment is in constant contraction, resulting in symptoms of bowel obstruction.1 Clinically, the symptoms of HD usually present immediately after birth (i.e. absence of meconium passage within the first 48 hours, vomiting, and abdominal distension), and as such, patients are often diagnosed in infancy. For older children, chronic constipation from birth and abdominal distension are classic symptoms of HD.2

Incidence of HD varies by geographical region, with rates ranging from 0.14 to 0.30 per 1,000 live births.3–7 Only one Canadian study has investigated the incidence of HD. A British Columbia surveillance cohort demonstrated incidence of 0.23
per 1,000 live births between 1964 and 1982. Incidence estimates are not available for Ontario, Canada’s most populous province. Recent studies have also reported an increasing incidence of HD in the general population. While this may be due to increased awareness of the disease and improved methods of detection, it must be noted that temporal trends have also been shown to vary by geographical region.

Ontario has a universal health care system in which all medically necessary direct health care costs (excluding medications) are paid by the provincial government for all legal residents (>99% of the population). These costs are contained within provincial health administrative data. These data represent an excellent opportunity to evaluate population-based estimates of incidence and outcomes of disease within the population. However, the accuracy of administrative data varies, and validation has been identified as a priority in the fields of epidemiological and health services research using these databases to minimize misclassification bias. This study used Ontario health administrative data, obtained using a validated algorithm, to determine the incidence and temporal trends of HD. Establishing a validated population-based cohort of HD patients will be invaluable in the future study of this condition, allowing for continued surveillance of identified patients.

Methods
This study was approved by the research ethics board of CHEO and the Ottawa Hospital.

Data sources
The health records of all legal Ontario residents (>99% of the population) are contained within anonymized provincial health administrative data, housed at the ICES. Each resident has a unique encrypted IKN based on his/her OHIP, allowing for deterministic linkage of a resident across health administrative and population databases. Investigators and analysts had access to uncleaned data from the full population of Ontario. We used the following datasets: hospitalization data from the CIHI-DAD, physician billing records from the OHIP database (including outpatient visits, emergency department care, and surgical procedures), population demographic data from the RPDB, and Canadian census data (census area profiles for 1991, 1996, 2001, 2006, and 2011). All entries within these databases are associated with a diagnostic code formatted to the ICD-9 before April 1, 2002 or ICD-10 after April 1, 2002.

Algorithm development and validation
To develop an algorithm for the identification of patients diagnosed with HD, true-positive (HD patients) and true-negative (patients without HD) reference standards were established. Potential true-positive cases of HD were identified within CHEO by two different methods. An electronic record search was conducted at CHEO between 1991 and 2010 to determine the true-positive reference standard (all patients <18 years of age diagnosed with HD at CHEO). CHEO is the only hospital within the CMA of Ottawa with inpatient pediatric beds or pediatric surgeries within the CMA of Ottawa. Therefore, all children with HD in this region are treated at this institution (i.e. HD is not a condition that would be treated at community or adult hospitals, unless diagnosis occurred at >18 years of age). The search for reference standard charts was performed using the ICD-9 and ICD-10 diagnostic codes for HD and other congenital functional disorders of the colon (ICD-9 751.3; ICD-10 Q43.1). The ICD code search was intended to be nonspecific and as inclusive as possible to ensure our true-positive cohort including all patients with potential HD. To minimize the potential for bias, an electronic search was conducted in the pathology database for the presence of the Systematized Nomenclature of Medicine Clinical Terms for biopsy associated with HD. All charts identified by these methods were reviewed by two reviewers (AN and a medical student) to confirm the diagnosis of HD using standard diagnostic criteria, and only patients born after April 1, 1988 residing in the CMA of Ottawa with a valid Ontario health card number were included. To establish a negative reference standard, the RPDB was used to identify all children <18 years of age living in Ottawa between 1991 and 2010 who were not identified by our search strategy and chart review, and therefore presumed not to have HD. This strategy has been shown to produce accurate true-negative reference standards in previous algorithm validation studies for the province of Ontario.

OHIP health card numbers for true-positive and true-negative reference standards were linked to the ICES-encrypted IKNs, allowing for the testing of various algorithms designed to identify HD patients in Ontario from within the health administrative data. We developed a total of 11 different algorithms using combinations of diagnostic and procedure codes from OHIP and the CIHI-DAD which had face validity for the identification of HD from within the data (Tables S1 and S2). We tested the suitability of each algorithm against the reference standards. We decided a priori that the algorithm that yielded the highest PPV, while maintaining a high sensitivity (optimally >90%), would be
selected as the one to be applied to the data to create the HD cohort. A higher PPV minimized false-positive identification of non-HD patients, and a higher sensitivity allowed for more complete identification of the cohort. This strategy has been used in the validation of algorithms for other rare diseases.12,14

Estimation of HD incidence in Ontario
The validated algorithm was applied to Ontario health administrative data to identify all HD cases in Ontario between 1991 and 2013. Inclusion criteria included hospital birth in Ontario between 1991 and 2013 with a valid health card number. Residents were excluded if they were not born in hospital, or if they migrated out of the province within the first year of life. Crude incidence of HD per 10,000 live births per fiscal year and overall was determined.

Statistical analysis
For the algorithm validation stage, we calculated the strength of each algorithm using the reference standards. We calculated sensitivity, specificity, PPV, and NPV with 95% CIs.

Crude incidence was calculated using the 2006 Canadian census standard population. Incidence time trends were assessed using sex-adjusted Poisson regression analysis. OR and 95% CIs were reported, with significance determined with a P-value of <0.05. To exclude patients with suspected short-segment HD from the evaluation of incidence trends, we conducted a sensitivity analysis to evaluate the trends in incidence in children diagnosed under 1 year of age separately from the overall cohort.

Results
Algorithm validation
To develop the true-positive reference standard, the charts of a total of 117 patients were screened, of which 41 were excluded due to birth before April 1, 1988 (n = 5) or the patients were not a resident of Ontario and thus did not have an OHIP number (n = 36) (Figure 1). A large number of non-Ontario residents were identified in the chart review as the catchment of CHEO includes Eastern Ontario and Western Quebec. The charts of all included patients were successfully linked to their health administrative data within the ICES database.

The ability of the 11 identification algorithms to correctly identify patients with HD varied widely (Table 1). The algorithm which identified patients with surgery/biopsy and hospitalization with HD as the true diagnosis (excluding diagnostic codes for suspected HD) was deemed to be the most accurate, and selected for utilization within the ICES database as it had the highest PPV (89.58%, 95% CI 77.34%–96.53%) and excellent sensitivity (93.48%, 95% CI 82.10%–98.63%).

Cohort creation and annual incidence estimates
By applying the validated algorithm to the administrative data, we identified a total of 679 patients <18 years of age diagnosed with HD in Ontario between 1991 and 2013. The majority of patients were male (75.41%, n = 512), living in an urban center (86.75%, n = 589), and had both rectal suction

Figure 1 Flow diagram of chart review process at CHEO. Patients diagnosed with HD (true-positive reference standard) were treated by corrective surgery using the Soave, Swenson, or Duhamel method.

Abbreviations: CHEO, Children’s Hospital of Eastern Ontario; HD, Hirschsprung disease.
biopsies and surgery (Table 2). The median age at diagnosis was 0.20 months (interquartile range: 0.07, 2.33 months).

The overall crude incidence rate for HD in Ontario between 1991 and 2013 was 2.05 per 10,000 live births (or 1 in 4,868 live births), with yearly values ranging from 0.98 per 10,000 to 3.08 per 10,000 live births (Figure 2 and Table S3). We observed no significant change in the incidence over time (OR 1.00, 95% CI 0.98–1.01, \( p = 0.80 \)). Sensitivity analysis to evaluate incidence in patients with long-segment disease (i.e. diagnosed under 1 year of age) indicated similar rates to the overall population (1.85 per 10,000 live births or 1 in 5,392; Figure S1).

**Discussion**

We have described the incidence and temporal trends of HD in Ontario, Canada, using validated population-based health administrative data. We determined that HD cases can be accurately identified from within health administrative data,

### Table 1 Algorithm validation

| Algorithm | Sensitivity (95% CI) | Specificity (95% CI) | PPV (95% CI) | NPV (95% CI) |
|-----------|----------------------|----------------------|-------------|-------------|
| 1 Hospitalization with HD in any field | 95.65 (85.16, 94.97) | >99.99 (99.99, 100.00) | 62.86 (50.48, 74.11) | >99.99 (99.99, 100.00) |
| 2 Hospitalization with HD in any field + surgery/biopsy | 93.48 (82.10, 98.63) | >99.99 (99.99, 100.00) | 84.31 (71.41, 92.98) | >99.99 (99.99, 100.00) |
| 3 Hospitalization with HD in any field + surgery (no biopsy) | 15.22 (6.34, 28.87) | >99.99 (99.99, 100.00) | 87.50 (47.35, 99.68) | >99.99 (99.99, 100.00) |
| 4 Hospitalization with HD in any field as TRUE diagnosis, do not include suspect/questionable diagnosis | 95.65 (85.16, 94.97) | >99.99 (99.99, 100.00) | 83.02 (70.20, 91.93) | >99.99 (99.99, 100.00) |
| 5 Hospitalization with HD in any field as TRUE diagnosis, include suspect/questionable diagnosis | 95.65 (85.16, 94.97) | >99.99 (99.99, 100.00) | 63.77 (51.31, 75.01) | >99.99 (99.99, 100.00) |
| 6 Hospitalization with HD in any field as TRUE diagnosis, do not include suspect/questionable diagnosis + surgery/biopsy | 93.48 (82.10, 98.63) | >99.99 (99.99, 100.00) | 89.58 (77.34, 96.53) | >99.99 (99.99, 100.00) |
| 7 Hospitalization with HD in any field as TRUE diagnosis, do not include suspect/questionable diagnosis + surgery (no biopsy) | 15.22 (6.34, 28.87) | >99.99 (99.99, 100.00) | 87.50 (47.35, 99.68) | >99.99 (99.99, 100.00) |
| 8 Any outpatient OHIP code (751) within first year of life | 58.70 (43.23, 73.00) | 99.78 (99.75, 99.81) | 10.80 (7.24, 15.32) | >99.99 (99.99, 100.00) |
| 9 Any outpatient OHIP code (751) within the first 2 years of life | 65.22 (49.75, 78.65) | 99.75 (99.72, 99.78) | 10.49 (7.19, 14.64) | >99.99 (99.99, 100.00) |
| 10 Any 2+ outpatient OHIP codes (751) within first year of life (on separate days) | 47.83 (32.89, 63.05) | 99.86 (99.84, 99.88) | 13.50 (8.66, 19.72) | >99.99 (99.99, 100.00) |
| 11 Any 2+ outpatient OHIP codes (751) within first 2 years of life (on separate days) | 58.70 (43.23, 73.00) | 99.84 (99.81, 99.86) | 14.06 (9.48, 19.80) | >99.99 (99.99, 100.00) |

**Notes:** 'Any hospitalization associated with ICD-9/10 code for HD in any field.' 'Any hospitalization associated with ICD-9/10 code for HD as “true” diagnosis.' The CIHI-DAD includes a “suspected” variable (INCLSUSPECT) which indicates that the diagnosis is suspected, not confirmed. ICES uses a macro that enables the algorithm to either include (INCLSUSPECT=T) or exclude (INCLSUSPECT=F) suspect/questionable diagnosis. Bold indicates the algorithm was that was applied to the data to create the final HD cohort.

**Abbreviations:** CI, confidence interval; CIHI-DAD, Canadian Institute for Health Information - Discharge Abstract Database; HD, Hirschsprung disease; ICD, International Classification of Diseases; ICES, Institute for Clinical Evaluative Sciences; NPV, negative predictive value; OHIP, Ontario Health Insurance Plan; PPV, positive predictive value.

### Table 2 General characteristics of the Ontario cohort (n = 679) identified as having HD by the selected algorithm

| Characteristic | N (%) |
|----------------|-------|
| Sex            |       |
| Male           | 512 (75.41) |
| Female         | 167 (24.59) |
| Age at diagnosis (years) |       |
| <1             | 613 (90.28) |
| 1–2            | 29 (4.27)  |
| 3–4            | 17 (2.50)  |
| ≥4             | 6 (0.88)   |
| Household at diagnosis |     |
| Rural          | 86 (12.67) |
| Urban          | 589 (86.75) |
| Rectal suction biopsies per patient |      |
| 0              | 140 (20.62) |
| 1              | 444 (65.39) |
| ≥2             | 77 (11.34)  |
| ≥3             | 11 (1.62)   |
| ≥4             | 7 (1.03)    |
| Intervention   |       |
| No surgery (biopsy only) | 86 (12.67) |
| Soave          | 100 (14.73) |
| Duhamel        | 88 (12.96)  |
| Other          | 405 (59.65) |

**Abbreviation:** HD, Hirschsprung disease.
and that incidence has not significantly changed in Ontario between 1991 and 2013.

Validation of an algorithm will allow us to continue surveillance of HD using Ontario data. While previous studies have utilized ICD codes to search health registries for cases of HD (including an earlier study from British Columbia, Canada3), this is the first study to validate the use of identification algorithms with health administrative data. Results from our study confirm the use of this method, with a sensitivity and PPV $\geq 90\%$. Similar methods were used to validate other disease cohorts within Ontario, yielding variable sensitivity and PPV measures. Our algorithm measures were similar to algorithms of ICD codes utilized in existing literature to establish incidence of intussusception (sensitivity 89.3\%, PPV 72.4\%),12 pediatric inflammatory bowel disease (sensitivity 89.6\%–90.5\%, PPV 59.2\%–76.0\%),14 pediatric asthma (sensitivity 91.4\%),15 and hospitalization of children for respiratory syncytial virus infection (sensitivity 97.9\%, PPV 96.9\%)16 within Ontario. A combination of procedural and diagnostic codes resulted in the most accurate identification of patients. While it is reasonable to assume that our algorithm may be used in other Canadian pediatric hospital based on the standardized training CIHI data entry personnel receive, our algorithm should be validated prior to application to the administrative data of other regions. For example, an Ontario study found that in the estimation of incidence of intussusception the addition of a procedural code to an algorithm of diagnostic codes dramatically reduced sensitivity.14 Conversely, a German study investigating incidence for the same condition found that the addition of a procedural code improved specificity while maintaining an acceptable sensitivity.17 Another Ontario study found that application of internationally validated algorithms to identify adults with inflammatory bowel disease had varying degrees of success in estimating inflammatory bowel disease in Ontario.18 This highlights the need to customize algorithms to the population and condition being examined, and to validate the algorithms against established reference standards prior to use.10

The incidence rate of HD in Ontario (1 in 4,868 live births) is similar to that most often reported in North America and Europe (1 in 5,000 live births).4,5,7,19–21 Our results are comparable to incidence rates described in British Columbia,3 Southeast Scotland,22 Denmark,23 and the USA24,25 (Table 3). This is not surprising given the proposed association between race and incidence rates for HD.8,24,26,27 While race and ethnicity are not available within Ontario health administrative data, the majority of the population of Ontario,28 British Columbia,28 Scotland,29 and Denmark30 are white. However, race/ethnicity could not account for all observations, such as Australia’s comparatively low incidence rate for HD,31 or the similarity in HD incidence between Ontario and Japan7,32 (Table 3). While these discrepancies may indicate the presence of additional factors yet to be uncovered in the etiology of HD, they may also be the result of study design. The Australian and Japanese studies estimated incidence rates based on self-reporting of surveyed clinicians and major hospitals, respectively. Further, these studies indicated less-than-optimal response rates, where only 81.1\% of Japanese
hospitals7 responded to their questionnaire and only 54% of Australian doctors completed the initial paper survey.31 Ultimately, all published incidence rates are estimates susceptible to any number of biases. This is supported by varying incidence rates observed in Denmark, despite the fact that both studies occurred in the same country during a similar time period.6,23

In addition to geographic differences, variations in temporal trends in HD incidence have been observed. Best et al showed a significant increase in incidence in North England between 1990 and 2008 ($p = 0.02$),4 and Koh et al also found a surge in cases in Tasmania between 2003 and 2005 for which no obvious explanation could be found.5 Contrary to these studies, our results did not show any evidence of an increasing trend in the incidence of HD in Ontario between 1991 and 2013. Incidence estimates from Baltimore,26 Japan,7 Denmark,6 and British Columbia3 also did not show a change in HD diagnosis across time. Without knowledge of the exact cause of HD, it is difficult to conclude why temporal trends are observed in some countries and not others. One hypothesis might be that incidence rates are increasing as a result of improvements in access to care or methods of diagnosis. In Ontario, where centralized surgical care was available to pediatric patients throughout the evaluation period (1991–2013), it is likely that access and investigative techniques did not change, resulting in stable incidence of HD. Ultimately, further research is required to assess the validity of a temporal trend and to determine what might be the cause for increased incidence rates of HD.

**Limitations**

The methodology used within our study to estimate the incidence rate of HD has strengths and weaknesses. Strengths include that estimates were made based on a population-based

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### Table 3 Literature review of estimates of HD

| Authors                        | Year                  | Location                  | Incidence rate |
|--------------------------------|-----------------------|---------------------------|----------------|
| Althoff22                       | 1945–1967             | Bremen, Germany           | 1/12,000       |
| Bodian and Carter34             | 1948–1959             | England                   | 1/2,000–1/10,000|
| Passarge25                      | 1948–1966             | Cincinnati, USA           | 1/5,000        |
| Orr and Scobie21                | 1953–1982             | Southeast Scotland        | 1/1,450        |
| Russell et al5                  | 1960–1964             | Denmark                   | 1/7,634        |
|                               | 1965–1969             |                           | 1/7,576        |
|                               | 1970–1974             |                           | 1/7,937        |
|                               | 1975–1979             |                           | 1/5,714        |
| Madsen23                       | Unclear (published in 1964) | Denmark                   | 1/4,700        |
| Spouge and Baird2               | 1964–1982             | British Columbia, Canada  | 1/4,417        |
| Goldberg26                      | 1969–1971             | Baltimore, USA            | 1/5,322        |
|                               | 1972–1974             |                           | 1/5,806        |
|                               | 1975–1977             |                           | 1/6,142        |
| Kleinhaus et al24               | 1975–1976             | USA                       | 1/5,257        |
| Ikeda and Goto22                | 1978–1982             | Japan                     | 1/4,697        |
| Suita et al7                   | 1978–1982             | Japan                     | 1/4,697        |
|                               | 1988–1992             |                           | 1/5,544        |
|                               | 1998–2002             |                           | 1/5,343        |
| Rajab et al4                   | 1989–1994             | Oman                      | 1/3,070        |
| Best et al4                    | 1990–1994             | North England             | 1/7,931        |
|                               | 1995–1999             |                           | 1/7,237        |
|                               | 2000–2004             |                           | 1/5,563        |
|                               | 2005–2008             |                           | 1/4,368        |
| Overall                        |                       |                           | 1/6,129        |
| Meza-Valencia et al21           | 1994–2002             | USA-associated Pacific Islands | 1/3,190      |
| Singh et al31                  | 1997–2000             | Australia                 | 1/7,165        |
| Koh et al5                     | 1998–2005             | Tasmania                  | 1/3,429        |
| Torfs27                        | Unclear (abstract published in 1998) | California                | White: 1/6,667 |
|                               |                       |                           | Black: 1/4,761 |
|                               |                       |                           | Hispanic: 1/10,000 |
|                               |                       |                           | Asian: 1/3,571 |

**Abbreviation:** HD, Hirschsprung disease.
incidence between 1991 and 2013. No change in incidence over time was comparable to previously published rates in Europe and North America, and no change in incidence over time was evident between 1991 and 2013.

Conclusion
Our study provided important information on the burden of HD in a large Canadian province. We described the creation of a population-based surveillance cohort of HD patients identified from within health administrative data using a validated algorithm. The estimated incidence of HD in Ontario was comparable to previously published rates in Europe and North America, and no change in incidence over time was evident between 1991 and 2013.

Abbreviations
CHEO, Children’s Hospital of Eastern Ontario; CI, confidence interval; CIHI-DAD, Canadian Institute for Health Information - Discharge Abstract Database; CMA, census metropolitan area; HD, Hirschsprung disease; ICD, International Classification of Diseases; ICES, Institute for Clinical Evaluative Sciences; IKN, identification number; NPV, negative predictive value; OHIP, Ontario HealthInsurance Plan; OR, odds ratio; PPV, positive predictive value; RPDB, registered persons database.

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Disclosure
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Supplementary materials

![Graph](image)

**Figure S1** Trends in crude incidence of HD in patients <1 year of age in Ontario over time.

**Abbreviation:** HD, Hirschsprung disease.

### Table S1 Diagnostic codes for HD from CIHI-DAD and OHIP

| Condition/procedure | Data source | Relevant entries |
|---------------------|-------------|------------------|
| **CIHI – diagnostic codes to identify HD** |             |                  |
| ICD-9 (1988–2001)   | Dxcode1–16  | 7513 = Hirschsprung disease |
| ICD-10 (2002–2013)  | Dscode1–25  | Q431 = Hirschsprung disease |
| **OHIP – diagnostic codes to identify HD** |             |                  |
| OHIP (1991–2013)    | Dxcode      | 751 = Hirschsprung megacolon, congenital malformation of the digestive system |

**Abbreviations:** CIHI-DAD, Canadian Institute for Health Information - Discharge Abstract Database; HD, Hirschsprung disease; ICD, International Classification of Diseases; OHIP, Ontario Health Insurance Plan.

### Table S2 Procedure codes related to HD from CIHI-DAD and OHIP

| Condition/procedure | Data source | Combination of codes |
|---------------------|-------------|----------------------|
| **CIHI – procedure codes** |             |                      |
| Soave               | Crcode1–20 (2002 onward) | 6031 = Soave submucosal resection of the rectum |
|                     | Prcode1–10 (1988–2001)   |                      |
|                     | Sprcode1–8 (1988–2001)   |                      |
| Duhamel             | Crcode1–20 (2002 onward) | 6054 = Duhamel resection |
|                     | Prcode1–10 (1988–2001)   |                      |
|                     | Sprcode1–8 (1988–2001)   |                      |
| Unspecified         | Crcode1–20 (2002 onward) | 603 = Pull-through resection of the rectum |
|                     | Prcode1–10 (1988–2001)   | 6039 = Other pull-through resection of the rectum |
|                     | Sprcode1–8 (1988–2001)   |                      |
| Miscellaneous       | Crcode1–20 (2002 onward) | 60 = Operations on rectum and perirectal tissue |
|                     | Prcode1–10 (1988–2001)   | 600 = Proctotomy     |
|                     | Sprcode1–8 (1988–2001)   | 601 = Proctostomy    |
|                     |                          | 602 = Local excision or destruction of lesion |
|                     |                          | 6021 = Fulguration of rectal lesion or tissue |
|                     |                          | 6022 = Destruction of rectal lesion or tissue B |
|                     |                          | 6023 = Destruction of rectal lesion or tissue B |
|                     |                          | 6024 = Local excision of rectal lesion or tissue |
|                     |                          | 604 = Abdominoperineal resection of rectum |
|                     |                          | 605 = Other resection of rectum |
|                     |                          | 6051 = Anterior resection with concomitant colon |
|                     |                          | 6052 = Other anterior resection |

(Continued)
### Table S2 (Continued)

| Condition/procedure | Data source | Combination of codes |
|---------------------|-------------|----------------------|
| 6053                |             | Posterior resection  |
| 6055                |             | Hartmann resection   |
| 6059                |             | Other resection of rectum NEC |
| 606                 |             | Repair of rectum    |
| 6061                |             | Suture of rectum    |
| 6062                |             | Closure of proctostomy |
| 6063                |             | Closure of other rectal fistula |
| 6064                |             | Rectorectostomy      |
| 6065                |             | Abdominal proctectomy|
| 6066                |             | Other proctectomy    |
| 6069                |             | Other repair of rectum |
| 607                 |             | Incision or excision of perirectal tissue |
| 6071                |             | Incision of perirectal tissue |
| 6072                |             | Excision of perirectal tissue |
| 6084                |             | Operative (transabdominal) proctosigmoid |
| 6089                |             | Other invasive diagnostic procedures on |
| 609                 |             | Other operations on rectum and perirectal |
| 6091                |             | Incision of rectal stricture |
| 6092                |             | Anorectal myectomy |
| 6093                |             | Repair of perirectal fistula |
| 6094                |             | Freeing of (intraluminal) adhesions of rectum |
| 6099                |             | Other operations on rectum and perirectal |

Incode miscellaneous 
Incode 1–20 (2002 onward)

1NQ87 = Excision partial, rectum
1NQ87BA = Excision partial, rectum endoscopic per orifice approach closure by apposition technique (e.g. suturing, stapling) or no closure required (for tissue regeneration)
1NQ87BFA = Excision partial, rectum endoscopic per orifice approach encirclement device
1NQ87CA = Excision partial, rectum perineal (e.g. pull through, transanal, sacral or sphincteric) approach closure by apposition technique (e.g. suturing, stapling) or no closure required (for tissue regeneration)
1NQ87DA = Excision partial, rectum endoscopic (laparoscopic, laparoscopic-assisted, hand-assisted) approach closure by apposition technique (e.g. suturing, stapling) or no closure required (for tissue regeneration)
1NQ87DE = Excision partial, rectum endoscopic (laparoscopic, laparoscopic-assisted, hand-assisted) approach colorectal anastomosis technique
1NQ87DF = Excision partial, rectum endoscopic (laparoscopic) approach colorectal anastomosis technique
1NQ87DX = Excision partial, rectum endoscopic (laparoscopic, laparoscopic-assisted, hand-assisted) approach stoma formation with distal closure
1NQ87LA = Excision partial, rectum open abdominal (e.g. anterior) approach closure by apposition technique (e.g. suturing, stapling) or no closure required (for tissue regeneration)
1NQ87PB = Excision partial, rectum perineal (e.g. pull through, transanal, sacral or sphincteric) approach colorectal anastomosis technique
1NQ87PF = Excision partial, rectum posterior (e.g. entering through incision between coccyx and anal verge with proctotomy) approach closure by apposition technique (e.g. suturing, stapling) or no closure required (for tissue regeneration)
1NQ87PN = Excision partial, rectum endoscopic (laparoscopic, laparoscopic-assisted, hand-assisted) approach robotic assisted telemanipulation of tools (telesurgery)
1NQ87RD = Excision partial, rectum open abdominal (e.g. anterior) approach colorectal anastomosis technique
1NQ87TF = Excision partial, rectum open abdominal approach (e.g. anterior) stoma formation with distal closure
1NQ89 = Excision total, rectum
1NQ89AB = Excision total, rectum, stoma formation with distal closure, combined endoscopic (laparoscopic) abdominoperineal approach

(Continued)
Table S2 (Continued)

| Condition/procedure | Data source       | Combination of codes                                                                                                                                 |
|---------------------|-------------------|-----------------------------------------------------------------------------------------------------------------------------------------------------|
|                     |                   | 1NQ89GV = Excision total, rectum combined endoscopic (abdominal) with perineal approach coloanal anastomosis technique                                 |
|                     |                   | 1NQ89KZ = Excision total, rectum abdominoperineal approach coloanal anastomosis technique                                                            |
|                     |                   | 1NQ89KZXXG = Excision total, rectum abdominoperineal approach pouch formation                                                                          |
|                     |                   | 1NQ89LH = Excision total, rectum abdominoperineal approach stoma formation with distal closure                                                        |
|                     |                   | 1NQ89LHXXG = Excision total, rectum abdominoperineal approach continent ileostomy formation                                                           |
|                     |                   | 1NQ89RS = Excision total, rectum abdominal (anterior) approach stoma formation with distal closure                                                    |
|                     |                   | 1NQ89RSXXG = Excision total, rectum abdominal (anterior) approach pouch formation                                                                       |
|                     |                   | 1NQ89SF = Excision total, rectum abdominal (anterior) approach coloanal anastomosis technique                                                              |
|                     |                   | 1NQ89SFXXG = Excision total, rectum abdominal (anterior) approach pouch formation                                                                         |
|                     |                   | 1NQ90 = Excision total with reconstruction, rectum                                                                                                       |
|                     |                   | 1NQ90LAXXXG = Excision total with reconstruction, rectum using open approach with ileum (for construction of pouch)                                       |
| Rectal suction biopsy | Prcode1–10 (1988–2001) | 6081 = Brush biopsy of rectum                                                                                                                       |
|                     |                   | 6082 = Other biopsy of rectum                                                                                                                       |
|                     |                   | 6083 = Biopsy of perirectal tissue                                                                                                                  |
|                     | Cprcode1–20 (2002 onward) | 608 = Invasive diagnostic procedures on rectum                                                                                                         |
|                     |                   | 6081 = Brush biopsy of rectum                                                                                                                       |
|                     |                   | 6082 = Other biopsy of rectum                                                                                                                       |
|                     | Incode1–20 (2002 onward) | 2NQ = Diagnostic interventions on the rectum                                                                                                          |
|                     |                   | 2NQ71 = Biopsy, rectum                                                                                                                            |
|                     |                   | 2NQ71BA = Biopsy, rectum using endoscopic per orifice approach                                                                                       |
|                     |                   | 2NQ71BG = Biopsy, rectum using endoscopic per orifice rectal suction                                                                                |
|                     |                   | 2NQ71BR = Biopsy, rectum using endoscopic per orifice with brush biopsy or washing                                                                  |
|                     |                   | 2NQ71CA = Biopsy, rectum per orifice approach NOS                                                                                                     |
|                     |                   | 2NQ71DA = Biopsy, rectum using endoscopic (laparoscopic) approach                                                                                     |
|                     |                   | 2NQ71HA = Biopsy, rectum using percutaneous (needle) approach (e.g. core needle biopsy)                                                                |
|                     |                   | 2NQ71LA = Biopsy, rectum using open approach                                                                                                          |

Notes: In 1988–2001 data, multiple variables contain required info (Prcode1–10, Sprcode1–8). If present, use only Prcode1–10 as the variables. Use Sprcode1–8 as a back-up if not present in Prcode1–10.

Abbreviations: CIHI-DAD, Canadian Institute for Health Information - Discharge Abstract Database; OHIP, Ontario Health Insurance Plan; NEC, not elsewhere classified; NOS, not otherwise specified.

Table S3 Annual crude incidence of HD in Ontario residents <18 years of age

| Year | Number of incident cases | Number of live births | Incidence per 10,000 live births | 95% LCL | 95% UCL |
|------|--------------------------|-----------------------|-------------------------------|--------|--------|
| 1991 | 17                       | 173,578               | 0.979                         | 0.571  | 1.568  |
| 1992 | 25                       | 157,367               | 1.589                         | 1.028  | 2.345  |
| 1993 | 29                       | 151,381               | 1.916                         | 1.283  | 2.751  |
| 1994 | 45                       | 151,724               | 2.966                         | 2.163  | 3.969  |
| 1995 | 27                       | 148,783               | 1.815                         | 1.196  | 2.64   |
| 1996 | 36                       | 141,785               | 2.539                         | 1.778  | 3.515  |
| 1997 | 24                       | 138,615               | 1.731                         | 1.109  | 2.576  |
| 1998 | 39                       | 136,481               | 2.858                         | 2.032  | 3.906  |
| 1999 | 42                       | 136,410               | 3.079                         | 2.219  | 4.162  |
| 2000 | 38                       | 132,472               | 2.869                         | 2.030  | 3.937  |
| 2001 | 28                       | 136,760               | 2.047                         | 1.360  | 2.959  |
| 2002 | 22                       | 135,087               | 1.629                         | 1.021  | 2.466  |

(Continued)
| Year | Number of incident cases | Number of live births | Incidence per 10,000 live births | 95% LCL | 95% UCL |
|------|--------------------------|-----------------------|----------------------------------|--------|--------|
| 2003 | 28                       | 139,765               | 2.003                            | 1.331  | 2.895  |
| 2004 | 29                       | 139,847               | 2.074                            | 1.389  | 2.978  |
| 2005 | 29                       | 141,087               | 2.055                            | 1.377  | 2.952  |
| 2006 | 28                       | 143,432               | 1.952                            | 1.297  | 2.821  |
| 2007 | 34                       | 146,653               | 2.318                            | 1.606  | 3.24   |
| 2008 | 22                       | 145,824               | 1.509                            | 0.945  | 2.284  |
| 2009 | 34                       | 135,629               | 2.507                            | 1.736  | 3.503  |
| 2010 | 40                       | 143,295               | 2.791                            | 1.994  | 3.801  |
| 2011 | 17                       | 144,440               | 1.177                            | 0.686  | 1.884  |
| 2012 | 24                       | 143,620               | 1.671                            | 1.071  | 2.486  |
| 2013 | 22                       | 141,380               | 1.556                            | 0.975  | 2.356  |

Abbreviations: HD, Hirschsprung disease; LCL, lower confidence limit; UCL, upper confidence limit.