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

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per 1,000 live births between 1964 and 1982.³ 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.^{5,8} 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.⁵⁻⁸

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.^{9,10} 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 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,¹¹ 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.^{12,13}

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.

Table 1 Algorithm validation

	Algorithm	Sensitivity (95% CI)	Specificity (95% CI)	PPV (95% CI)	NPV (95% CI)
1	Hospitalization with HD in any field ^a	95.65 (85.16, 99.47)	>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 ^a + 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 ^a + 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 ^b , do not include suspect/questionable diagnosis ^c	95.65 (85.16, 99.47)	>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 ^b , include suspect/questionable diagnosis ^c	95.65 (85.16, 99.47)	>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^b, do not include suspect/questionable diagnosis^c + 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 ^b , do not include suspect/questionable diagnosis ^c + 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: ^aAny hospitalization associated with ICD-9/10 code for HD in any field. ^bAny hospitalization associated with ICD-9/10 code for HD as “true” diagnosis. ^cThe CIHI-DAD includes a “suspected” variable (INCLUSUSPECT) which indicates that the diagnosis is suspected, not confirmed. ICES uses a macro that enables the algorithm to either include (INCLUSUSPECT=T) or exclude (INCLUSUSPECT=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.

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 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)
	2–3	17 (2.50)
	3–4	6 (0.88)
	≥4	14 (2.06)
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.

Figure 2 Trends in crude incidence of HD in patients <18 years of age in Ontario over time.
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 USA.^{24,25} (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 Japan.^{7,32} (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

Table 3 Literature review of estimates of HD

Authors	Year	Location	Incidence rate
Althoff ³³	1945–1967	Bremen, Germany	1/12,000
Bodian and Carter ³⁴	1948–1959	England	1/2,000–1/10,000
Passarge ²⁵	1948–1966	Cincinnati, USA	1/5,000
Orr and Scobie ²²	1953–1982	Southeast Scotland	1/1,450
Russell et al ⁶	1960–1964	Denmark	1/7,634
	1965–1969		1/7,576
	1970–1974		1/7,937
	1975–1979		1/5,714
			Overall = 1/7,165
Madsen ²³	Unclear (published in 1964)	Denmark	1/4,700
Spouge and Baird ³	1964–1982	British Columbia, Canada	1/4,417
Goldberg ²⁶	1969–1971	Baltimore, USA	1/5,322
	1972–1974		1/5,806
	1975–1977		1/6,142
			Overall = 1/5,692
Kleinhaus et al ²⁴	1975–1976	USA	1/5,257
Ikeda and Goto ³²	1978–1982	Japan	1/4,697
Suita et al ⁷	1978–1982	Japan	1/4,697
	1988–1992		1/5,544
	1998–2002		1/5,343
Rajab et al ⁴	1989–1994	Oman	1/3,070
Best et al ⁸	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 al ²¹	1994–2002	USA-associated Pacific Islands	1/3,190
Singh et al ³¹	1997–2000	Australia	1/7,165
Koh et al ⁵	1998–2005	Tasmania	1/3,429
Torfs ²⁷	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.

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 ($p = 0.02$),⁸ 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

cohort rather than a smaller subset, and were therefore not subject to ascertainment bias. Our validation of the algorithms used to identify HD patients is an additional strength, although the possibility of misclassification bias (occurring both in the identification of the reference standard and the population cohort) can never be excluded in studies using health administrative data. For example, it is thought that misclassification might have contributed to some of our more unusual results, including 12.67% of patients having no surgery and over 50% of patients receiving “other” intervention in our population. A similar concern is a lack of coding for minor procedures, such as biopsy, which surgeons often waive when billing (potentially supported by the 20.62% of patients without biopsy in Table 2). In addition, very mild patients with short-segment HD who may have presented in adulthood with chronic constipation would not have been identified by our algorithm. The derivation of our algorithm from a single cohort that was not validated outside of our center (due to feasibility and budget limitations) may also represent a weakness of our study. However, CHEO represents the only pediatric hospital in the region, and is therefore representative of care received by children in the entire region of Eastern Ontario. In addition, the final algorithm was based only on discharge data (DAD) and surgical codes from the CIHI. CIHI is a national organization tasked with training professional coders in all hospitals on accurate coding, and thus, all pediatric hospital coders receive the same training in Ontario. Therefore, we have reason to believe that an algorithm validated in one institution would perform adequately in other institutions in the province. However, we acknowledge that the ability of our algorithm to identify HD cases could vary across jurisdictions depending on practice variations in hospitalization.

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 Health Insurance Plan; OR, odds ratio; PPV, positive predictive value; RPDB, registered persons database.

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Disclosure

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Supplementary materials

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)	DxcodeI–I6	7513 = Hirschsprung disease
ICD-10 (2002–2013)	DxI0codeI–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	CprcodeI–20 (2002 onward) PrcodeI–10 (1988–2001) SprcodeI–8 (1988–2001)	6031 = Soave submucosal resection of the rectum
Duhamel	CprcodeI–20 (2002 onward) PrcodeI–10 (1988–2001) SprcodeI–8 (1988–2001)	6054 = Duhamel resection
Unspecified	CprcodeI–20 (2002 onward) PrcodeI–10 (1988–2001) SprcodeI–8 (1988–2001)	603 = Pull-through resection of the rectum 6039 = Other pull-through resection of the rectum
Miscellaneous	CprcodeI–20 (2002 onward) PrcodeI–10 (1988–2001) SprcodeI–8 (1988–2001)	60 = Operations on rectum and perirectal tissue 600 = Proctotomy 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
Incode miscellaneous	IncodeI–20 (2002 onward)	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 proctopexy
		6066 = Other proctopexy
		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
		INQ87 = Excision partial, rectum
		INQ87BA = Excision partial, rectum endoscopic per orifice approach closure by apposition technique (e.g. suturing, stapling) or no closure required (for tissue regeneration)
		INQ87BAFA = Excision partial, rectum endoscopic per orifice approach encircage device
		INQ87CA = 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)
		INQ87DA = 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)
		INQ87DE = Excision partial, rectum endoscopic (laparoscopic, laparoscopic-assisted, hand-assisted) approach colorectal anastomosis technique
		INQ87DF = Excision partial, rectum endoscopic (laparoscopic) approach colorectal anastomosis technique
		INQ87DX = Excision partial, rectum endoscopic (laparoscopic, laparoscopic-assisted, hand-assisted) approach stoma formation with distal closure
		INQ87LA = Excision partial, rectum open abdominal (e.g. anterior) approach closure by apposition technique (e.g. suturing, stapling) or no closure required (for tissue regeneration)
		INQ87PB = Excision partial, rectum perineal (e.g. pull through, transanal, sacral or sphincteric) approach colorectal anastomosis technique
		INQ87PF = 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)
		INQ87PN = Excision partial, rectum endoscopic (laparoscopic, laparoscopic-assisted, hand-assisted) approach robotic assisted telemanipulation of tools (telesurgery)
		INQ87RD = Excision partial, rectum open abdominal (e.g. anterior) approach colorectal anastomosis technique
		INQ87TF = Excision partial, rectum open abdominal approach (e.g. anterior) stoma formation with distal closure
		INQ89 = Excision total, rectum
		INQ89AB = Excision total, rectum, stoma formation with distal closure, combined endoscopic (laparoscopic) abdominoperineal approach

(Continued)

Table S2 (Continued)

Condition/procedure	Data source	Combination of codes
		INQ89GV = Excision total, rectum combined endoscopic (abdominal) with perineal approach coloanal anastomosis technique
		INQ89KZ = Excision total, rectum abdominoperineal approach coloanal anastomosis technique
		INQ89KZXXG = Excision total, rectum abdominoperineal approach pouch formation
		INQ89LH = Excision total, rectum abdominoperineal approach stoma formation with distal closure
		INQ89LHXXG = Excision total, rectum abdominoperineal approach continent ileostomy formation
		INQ89RS = Excision total, rectum abdominal (anterior) approach stoma formation with distal closure
		INQ89RSXXG = Excision total, rectum abdominal (anterior) approach continent ileostomy formation
		INQ89SF = Excision total, rectum abdominal (anterior) approach coloanal anastomosis technique
		INQ89SFXXG = Excision total, rectum abdominal (anterior) approach pouch formation
		INQ90 = Excision total with reconstruction, rectum
		INQ90LAXXG = Excision total with reconstruction, rectum using open approach with ileum (for construction of pouch)
Rectal suction biopsy	PrccodeI-10 (1988-2001)	6081 = Brush biopsy of rectum
		6082 = Other biopsy of rectum
		6083 = Biopsy of perirectal tissue
	CprccodeI-20 (2002 onward)	608 = Invasive diagnostic procedures on rectum
		6081 = Brush biopsy of rectum
		6082 = Other biopsy of rectum
	InccodeI-20 (2002 onward)	2NQ = Diagnostic interventions on the rectum
		2NQ7I = Biopsy, rectum
		2NQ7IBA = Biopsy, rectum using endoscopic per orifice approach
		2NQ7IBG = Biopsy, rectum using endoscopic per orifice rectal suction
		2NQ7IBR = Biopsy, rectum using endoscopic per orifice with brush biopsy or washing
		2NQ7ICA = Biopsy, rectum per orifice approach NOS
		2NQ7IDA = Biopsy, rectum using endoscopic (laparoscopic) approach
		2NQ7IHA = Biopsy, rectum using percutaneous (needle) approach (e.g. core needle biopsy)
		2NQ7ILA = Biopsy, rectum using open approach

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

Table S3 (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.

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