Huntington Disease in Canada

Methodology Notes

Canadian Institute for Health Information
Institut canadien d'information sur la santé

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Identifying information

Project name	Huntington Disease (HD) in Canada
Project description	The Canadian Institute for Health Information (CIHI) has received funding from the Health Canada–led National Strategy for Drugs for Rare Diseases to improve the collection and use of real-world evidence to support decision-making. This project explores and reports how the rare disease Huntington disease (HD) is captured in databases at CIHI; this project also provides baseline information about the portrait of care for patients with adult HD. This analysis includes 5 parts: 1. Identifying residents with adult HD in long-term care (LTC) 2. Describing care, demographics and health characteristics of adult HD in LTC facilities 3. Understanding prescription drug use among LTC residents with adult HD 4. Understanding prescription drug use among all LTC residents 5. Describing the patient population and care for adult HD in outpatient clinics
Project time frame	November 2023 to November 2024
Resources	*Indicator Library: General Methodology Notes – Clinical Indicators* Continuing Care Metadata Integrated interRAI Reporting System (IRRS) metadata National Prescription Drug Utilization Information System (NPDUIS) metadata Pharmaceutical Data Tool National Ambulatory Care Reporting System (NACRS) metadata
General inquiries	drugs@cihi.ca

Project analysis 1: Adult HD LTC cohorts

Objective of analysis	To identify residents with adult HD in long-term care (LTC) facilities
Data sources	Continuing Care Reporting System (CCRS), CIHI: The CCRS data standard is the Resident Assessment Instrument–Minimum Data Set 2.0 (RAI-MDS 2.0) Integrated interRAI Reporting System (IRRS), CIHI: The IRRS data standard for LTC is the interRAI Long-Term Care Facilities (interRAI LTCF)
Data time frame	Fiscal years 2020–2021 to 2021–2022
Geographic coverage	CCRS: Newfoundland and Labrador, Ontario, Alberta, British Columbia IRRS interRAI LTCF: New Brunswick, Saskatchewan (see Appendix A for details)
Cohort description	1. Cohort 1 (existing LTC residents with adult HD): Residents age 21 years and older with an LTC assessment and an HD diagnosis in 2021–2022 2. Cohort 2 (newly admitted LTC residents with adult HD): Residents age 21 years and older with an LTC assessment and an HD diagnosis with an admission date in 2021–2022
Inclusions	**Cohorts 1 and 2** 1. Assessments between April 1, 2021, and March 31, 2022 2. LTC assessments only 3. LTC residents with an HD diagnosis i. In CCRS, the disease diagnoses pick-list (data element I1X_Huntingtons_Chorea) was used to identify HD. ii. In IRRS interRAI LTCF, the ICD-10-CA codes G10, G308.0 or F02.2 (present in any of the Other Disease Diagnoses fields) were used to identify HD. Note: At the time of analysis, the IRRS interRAI LTCF instrument did not include HD in its disease diagnoses pick-list. 4. Age at entry greater than or equal to 21 years 5. Gender equal to male or female **Cohort 2** 1. LTC admission date between April 1, 2021, and March 31, 2022
Exclusions	1. Cohorts 1 and 2: LTC residents with missing or invalid encrypted health card number or health card issuing province. 2. Cohort 2: For each resident, a 1-year look-back period from the date of admission to LTC in 2021–2022 was used to identify prior LTC assessments with HD recorded; residents with an assessment in this period were excluded.
Linkage	Assessment and episode data were linked using the data element episode_id

Project analysis 2: Care and demographic characteristics of LTC residents with adult HD

Objective of analysis	To describe care, demographic and health characteristics of long-term care (LTC) residents with adult HD
Data sources	Continuing Care Reporting System (CCRS), CIHI: The CCRS data standard is the Resident Assessment Instrument–Minimum Data Set 2.0 (RAI-MDS 2.0) Integrated interRAI Reporting System (IRRS), CIHI: The IRRS data standard for LTC is the interRAI Long-Term Care Facilities (interRAI LTCF)
Data time frame	Fiscal years 2021–2022 to 2022–2023
Cohort description	Existing (Cohort 1) and newly admitted (Cohort 2) LTC residents with adult HD (see Project analysis 1)
Calculation description	**LTC residents with HD profile (Cohort 1)** Note: Calculations are based on a resident's last assessment, all assessments or their first assessment in 2021–2022, as noted below. Metrics correspond to data elements from the LTC assessment instruments, unless otherwise specified. 1. LTC residents with HD i. The total number of residents with at least 1 assessment with an HD diagnosis, based on all assessments in 2021–2022 2. Province of residence of LTC residents with HD i. The number and percentage of LTC residents with HD by province, based on all assessments in 2021–2022 3. Age of LTC residents with HD i. The number and percentage of LTC residents with HD in each age group (21–49, 50–59, 60–69, 70–79 and 80+), calculated using age at their last assessment in 2021–2022 ii. The mean, median and age range of LTC residents with HD, based on their last assessment in 2021–2022 4. Recorded sex or gender of LTC residents with HD i. The number and percentage of female and male LTC residents with HD, based on their last assessment in 2021–2022 5. Urban versus rural/remote i. The number and percentage of LTC residents with HD in urban or rural/remote residence categories, based on their last assessment in 2021–2022 LTC residents with HD were assigned to urban or rural/remote communities, based on the postal codes of place of residence using the Postal Code Conversion File Plus (PCCF+) version 8A. Residents with unknown postal codes were excluded. 6. Neighbourhood income quintile i. The number and percentage of LTC residents with HD in each neighbourhood income quintile, based on their last assessment in 2021–2022 LTC residents with HD were assigned to a neighbourhood income quintile based on the postal codes of place of residence using the PCCF+ version 8A. Residents whose neighbourhood income quintile was unknown were excluded.

Calculation description (continued)	7. Source of admission i. The type of residence and the specific facility/level of care the LTC resident with HD was admitted from, as recorded in the episode 8. Discharge disposition, based on the latest available data at the time of analysis i. The total number of residents with a record of discharge (as of March 31, 2024) ii. The location where the LTC resident with HD was discharged to, or the status of the resident with HD on discharge based on their last assessment (as of March 31, 2024) iii. *Died in facility* includes deaths between April 1, 2021, and March 31, 2024 9. Length of stay i. The average length of stay of LTC residents with HD, calculated as the number of days from admission date to the last assessment date in 2021–2022 10. Disease diagnoses i. The number and percentage of LTC residents with HD with a diagnosis of anxiety, bipolar disorder, depression, pneumonia, schizophrenia and/or seizure disorder, based on all assessments in 2021–2022 11. Health conditions i. The number and percentage of LTC residents with a fracture, fall (in the last 30 days), recurrent lung aspiration (in the last 90 days), unsteady gait and/or urinary tract infection (in the last 30 days), based on all assessments in 2021–2022 12. Continence (bowel and bladder) i. The number and percentage of LTC residents with HD within each level of bladder and bowel incontinence, based on their last assessment in 2021–2022 13. Oral and nutritional status, based on all assessments in 2021–2022 i. The number and percentage of LTC residents with HD with a chewing problem, feeding tube, oral feeding (syringe), parenteral IV, swallowing problem and/or weight loss (last 180 days) ii. The number and percentage of LTC residents with HD using dietary supplements and/or on a planned weight change program, based on CCRS data only 14. Therapies, based on all assessments in 2021–2022 i. The number and percentage of LTC residents with HD who received any of the following types of therapy: speech, psychological, recreational, occupational, physical and/or respiratory ii. The number and percentage of LTC residents with HD with speech, psychological, recreational, occupational, physical and/or respiratory therapy 15. Additional metrics, based on all assessments in 2021–2022 i. The number and percentage of LTC residents with HD who use a cane, walker or crutch as their primary mode of locomotion ii. The number and percentage of LTC residents with HD who use a wheelchair as their primary mode of locomotion iii. The number and percentage of LTC residents with HD who had unclear or no speech iv. The number and percentage of LTC residents with HD who lived alone prior to entry into LTC

Calculation description (continued)	16. Age at death, based on most recent data available at the time of analysis 17. The age distribution (22–49, 50–59, 60–69, 70–79 and 80+) of LTC residents with HD who died based on the age at assessment for assessments with a discharge recorded as death from April 1, 2021, to March 31, 2024 **Outcome scales, based on each resident's last assessment in 2021–2022 (Cohort 1)** 1. The distribution of the categorical score for the following scales, including the number and percentage of LTC residents with HD within each category: i. Cognitive Performance Scale (CPS) ii. Activities of Daily Living (ADL) Self-Performance Hierarchy Scale iii. Depression Rating Scale iv. Aggressive Behaviour Scale v. Index of Social Engagement (based on CCRS data only) vi. Pain Scale **Metrics for newly admitted LTC residents with adult HD (Cohort 2)** 1. The above metrics were also generated for LTC residents with adult HD admitted to LTC in 2021–2022, except for discharge disposition and forms of therapy (many missing values). Metrics were based on each resident's first assessment with HD in 2021–2022. **Metrics by cognitive status** 1. The above metrics were also generated by cognitive status (intact or impaired) of LTC residents with HD except for forms of therapy (many missing values). Using Cohort 1, cognitive status was determined based on each resident's last assessment in 2021–2022. Residents with a CPS categorical score of "intact" or "borderline intact" were grouped into the "intact cognition" group, while those with scores of "mild" to "very severe" impairment were grouped into the "impaired cognition" group.

Project analysis 3: Prescription drug use of LTC residents with adult HD

Objective of analysis	To describe prescription drug use of long-term care (LTC) residents with adult HD
Data sources	National Prescription Drug Utilization Information System (NPDUIS), CIHI For information on plans and programs included in NPDUIS, see Appendix B.
Data time frame	Fiscal years 2021–2022 to 2022–2023
Geographic coverage	Newfoundland and Labrador, New Brunswick, Ontario, Saskatchewan, British Columbia (see Appendix B for details)
Cohort description	Existing LTC residents with adult HD (Cohort 1) were linked to their drug claims.
Inclusions	Residents with at least 1 drug claim submitted to a public drug program for payment or for processing for documentation under a drug information system (DIS). Drug claims over a 1-year period from each LTC resident's first assessment in 2021–2022 were included in this analysis.
Exclusions	Alberta (see Appendix B for details)
Linkage	Claims data from NPDUIS was linked to Cohort 1 using CIHI's standard client linkage methodology. A linking key was created by combining the data elements Province/Territory Issuing Health Card Number and Health Card Province Code.
Calculation description	1. The number of drug classes per LTC resident with HD, all claims* i. The unique number of drug classes, defined by the World Health Organization (WHO) anatomical therapeutic chemical (ATC) level 4 code, prescribed per LTC resident with HD in a 1-year period, grouped into 4 categories: 1 to 4, 5 to 9, 10 to 14, and 15 or more Non-drug products and products without an assigned ATC code were excluded from this analysis. Non-drug products include, but are not limited to, diabetes supplies, wound care, ostomy supplies and pharmaceutical services. 2. The types of drugs prescribed to LTC residents with HD, public claims only† i. The percentage of claims for generic, brand name and biologic drug products, calculated as the number of claims for each drug type divided by the total number of claims ii. The percentage of spending for generic, brand name and biologic drug products, calculated as the spending for each drug type divided by the total program spending Over-the-counter and non-drug products were excluded from this analysis.

Calculation description (continued)	3. Drugs prescribed to LTC residents with HD by ATC level 3, 4 and 5 groups, all claims* i. Rate of use Calculated as the number of LTC residents with HD with at least 1 claim for a given ATC group over the total number of LTC residents with HD with at least 1 claim. ii. The number of LTC residents with HD with at least 1 claim for a given ATC group Non-drug products and products without an assigned ATC code were excluded from this analysis. 4. Costs of drugs prescribed to LTC residents with HD, public claims only† i. Total program paid amount: The amount from the total prescription cost accepted that is paid by the plan/program. This amount includes the drug cost as well as the associated professional fee and markup, if applicable. ii. Total cost claimed amount: The total dollar amount of a prescription claimed to the plan/program, as it relates to quantity claimed. This amount includes the drug cost as well as the associated professional fee and markup, if applicable. 5. Costs of drugs prescribed to LTC residents with HD by ATC level 5 group, public claims only† i. Total program spending ii. Proportion of total program spending Calculated as the program spending for a given chemical, divided by the total program spending for all public drug claims. iii. Rate of use Calculated as the number of LTC residents with HD with at least 1 public drug claim for a given chemical, divided by the total number of LTC residents with HD with at least one public drug claim. iv. The number of LTC residents with HD with at least 1 public drug claim for a given chemical **Notes** * *All claims* are claims dispensed in community pharmacies (i.e., public and private claims). These claims include those submitted to public drug programs for payment or for processing for documentation under a DIS. † *Public claims* are claims where at least part of the claim was submitted to the public plan/program. *ATC level* is a classification system that divides drugs into different groups according to the organ or system on which they act and their chemical, pharmacological and therapeutic properties. This analysis used the 2024 version of the WHO ATC classification system.

Project analysis 4: Prescription drug use among all LTC residents

Objective of analysis	To describe prescription drug use among all long-term care (LTC) residents (comparison population for Project analysis 3)
Data sources	National Prescription Drug Utilization Information System (NPDUIS), CIHI For information on plans and programs included in NPDUIS, see Appendix B.
Data time frame	Fiscal year 2021–2022
Geographic coverage	Provinces with LTC-identifiable drug claims in NPDUIS: Newfoundland and Labrador, New Brunswick, Ontario, British Columbia (see Appendix B for details)
Cohort description	All residents in LTC facilities (includes LTC residents from most provinces included in Project analysis 3)
Inclusions	Drug claims submitted to public LTC drug programs for payment
Exclusions	Saskatchewan, Alberta (see Appendix B for details)
Linkage	None
Calculation description	1. The number of drug classes prescribed per LTC resident, all claims* i. The unique number of drug classes (ATC level 4 code) prescribed per LTC resident in 2021–2022, grouped into 4 categories: 1 to 4, 5 to 9, 10 to 14, and 15 or more Non-drug products and products without an assigned ATC code were excluded from this analysis. Non-drug products include, but are not limited to, diabetes supplies, wound care, ostomy supplies and pharmaceutical services. Over-the-counter and non-drug products were excluded from this analysis. 2. Drugs prescribed to LTC residents by ATC level 3, all claims* i. The rate of use Calculated as the number of LTC residents with at least one claim for a given ATC level 3 group over the total number of LTC residents. Non-drug products and products without an assigned ATC code were excluded from this analysis. **Notes** * *All claims* are claims dispensed in community pharmacies (i.e., public and private claims). These claims include those submitted to public drug programs for payment or for processing for documentation under a DIS. *ATC level* is a classification system that divides drugs into different groups according to the organ or system on which they act and their chemical, pharmacological and therapeutic properties. This analysis used the 2024 version of the WHO ATC classification system.

Project analysis 5: Clinic care for patients with adult HD

Objective of analysis	To describe clinic care for patients with adult HD
Data sources	National Ambulatory Care Reporting System (NACRS), CIHI
Data time frame	Fiscal year 2022–2023
Geographic coverage	Alberta Note: Reporting of clinic data to CIHI was interrupted by implementation of a new clinical information reporting system in Alberta, Connect Care, in 2019. As a result, some clinics no longer submit data to NACRS. NACRS clinic data for 2022–2023 reflects partial submission. There were 20% fewer patients with HD in 2022–2023 compared to the full data submission period, with no differences in demographics.
Cohort description	Patients age 21 years and older who visited a clinic and had an HD diagnosis
Inclusions	1. Patients with a clinic visit (amcare code = CL) between April 1, 2022, and March 31, 2023 2. HD diagnosis code (ICD-10-CA: G10, G308.0, F02.2) in any diagnosis code position 3. Age 21 years old or older at the date of registration 4. Facility province in Alberta 5. Patients who had a submitting province code of Alberta
Exclusions	Invalid encrypted health card number or health card issuing province
Calculation description	**Patient profile** 1. Patients with HD i. The number of unique patients who had at least one clinic visit with an HD diagnosis 2. Recorded sex or gender of patients i. The number and percentage of female and male (as documented in the health record) patients with HD with a clinic visit 3. Location of residence i. The number and percentage of patients in urban or rural/remote residence categories Patients were assigned to urban or rural/remote communities based on the postal codes of place of residence using the PCCF+ version 8A. Patients with unknown postal codes were excluded. 4. Neighbourhood income quintile i. The number and percentage of patients in each neighbourhood income quintile Patients were assigned to a neighbourhood income quintile based on the postal codes of place of residence using the PCCF+ version 8A. Patients with unknown neighbourhood income quintiles were excluded.

Calculation description (continued)	5. Age of patients (numeric) i. The age distribution (mean, minimum, maximum, 10th, 25th, 50th, 75th and 90th percentiles) of the cohort Age as recorded on the health record on the date of registration **Care utilization** 1. Clinic visits i. The total number of visits ii. The distribution (mean, minimum, maximum, standard deviation, 10th, 25th, 50th, 75th and 90th percentiles) of the number of visits per patient iii. The number of clinic visits per patient and the percentage of the cohort with the following number of visits: 1, 2, 3, 4–7 or 10+ 2. Clinic type and provider characteristics i. The number and percentage of patients by clinic type ii. The number and percentage of visits by main provider specialty 3. Mode of visit i. The number and percentage of visits by mode (in person or by telephone or videoconference), all patients ii. The number and percentage of visits by mode for patients younger than age 65 and age 65 and older 4. Time and distance travelled for clinic care i. The number and percentage of patients who travelled less than 20 km, 20 to 49 km, 50 to 99 km and 100 km or more to their clinic visit ii. The number and percentage of patients who travelled less than 15 min, 15 to 29 min, 30 to 59 min and 60 min or more to their clinic visit To calculate an estimated distance from patient residence to treatment facility, a geographic information system (GIS) was used. Esri's ArcGIS Pro desktop platform, in conjunction with the Network Analyst extension, was used with Statistics Canada's *Road Network File, 2018* (geospatial data layer). Patient residence locations were approximated based on postal code, with latitude and longitude coordinates derived using Statistics Canada's PCCF+ version 8A. Treatment facility locations were also derived using postal codes and the PCCF+ version 8A.

Quality statement

Exceptions and limitations	**Cell suppression methodology for all requested data tables** In accordance with CIHI's privacy policy, when the cell size is less than 5 but greater than 0, this number and its associated metrics are suppressed to ensure confidentiality. When the difference in the numbers between corresponding entries (rows) in different tables is less than 5, the number and other associated values are also suppressed to avoid residual disclosure. Note: There may be exceptions to this rule if warranted. **Long-term care (LTC) reporting period** Since project analysis 3 involved analyzing drug claims prescribed to LTC residents over a 1-year period following their first assessment, 2021–2022 data was used to identify the LTC cohorts. At the time of analysis, drug data was available up to fall 2023. Since the LTC analysis uses data from two different reporting instruments, Continuing Care Reporting System (CCRS) and Integrated interRAI Reporting System (IRRS), data elements may not be comparable or present in both data sets. Our analysis includes data elements that are comparable and some that are present only in CCRS, as described in each methodologic summary above. **NPDUIS** Due to the design of public drug programs in Canada (i.e., seniors and low-income families/individuals are the only populations covered in all jurisdictions), CIHI has limited data on claims made by non-seniors. As a result, NPDUIS is not a population-based system (except for Manitoba, Saskatchewan and British Columbia). There may also be differences in population characteristics (such as age and health status) between seniors with and without public coverage. In provinces where a lower proportion of seniors have claims accepted by the public plan (e.g., Newfoundland and Labrador, Nova Scotia, New Brunswick), drug utilization patterns among those with public coverage are more likely to be affected by these differences and, therefore, may be less reflective of utilization patterns among all seniors in the province. Claims for drugs dispensed in hospitals, as well as for those funded through cancer agencies, are not submitted to NPDUIS. NPDUIS does not include information regarding • Prescriptions that were written but never dispensed; • Prescriptions that were dispensed but for which the associated drug costs were not submitted to, or not accepted by, the public drug programs; or • Diagnoses or conditions for which prescriptions were written.

Appendices

Appendix A: Notes for LTC analysis

Geographic inclusions

Newfoundland and Labrador, New Brunswick, Ontario, Saskatchewan, Alberta and British Columbia have full coverage of LTC data in either CCRS or IRRS in 2021–2022.

The Yukon has full coverage in CCRS but was excluded due to very low (<5) patient counts. Prince Edward Island, Quebec, the Northwest Territories and Nunavut were excluded since they do not submit any LTC data to CCRS or IRRS. Nova Scotia and Manitoba have limited coverage (for only 1 facility and from the Winnipeg Regional Health Authority, respectively) and were therefore also excluded.

Appendix B: Notes for linked drug analysis

Table B1 Plans/programs in the NPDUIS

Jurisdiction	Plan/program description
Newfoundland and Labrador	• 65Plus Plan • Access Plan • Assurance Plan • Foundation Plan • Pandemic Plan • Select Needs/Cystic Fibrosis Plan • Select Needs/Growth Hormone Plan
New Brunswick	• Extra-Mural Program • Medical Abortion Program • New Brunswick Drug Plan • New Brunswick Prescription Drug Program – Children in Care of the Minister Social Development and Special Needs Children – Cystic Fibrosis – Growth Hormone Deficiency – HIV/AIDS – Individuals in Licensed Residential Facilities – Multiple Sclerosis – Nursing Home Residents – Organ Transplant Recipients – Seniors – Social Development Clients • Pharmacy Administered Publicly Funded Vaccines, Testing and Drug Therapies Program • Tuberculosis Drug Plan
Ontario	• Ministry of Children, Community and Social Services (MCCSS) • Ontario Drug Benefit Program (ODB)
Saskatchewan	• Universal Program • Non-adjudicated

Jurisdiction	Plan/program description
British Columbia	• Assurance • Children in the At Home Program • Cystic Fibrosis • Fair PharmaCare • Nicotine Replacement Therapies • Palliative Care • Psychiatric Medication Program • Recipients of B.C. Income Assistance • Residential Care • Non-adjudicated

For more information regarding plans and programs submitted in NPDUIS, refer to the Pharmaceutical Data Tool.

Geographic inclusions

The provinces included in the LTC analysis were eligible for inclusion in the linked drug analysis. LTC drug claims data from Alberta is not submitted to NPDUIS. Prescription drug data for Saskatchewan and British Columbia includes both public and private drug claims. Ontario data includes public drug claims; however, since the Ontario Drug Benefit program is the first payer and covers residents in LTC, data coverage was assumed to be relatively complete. Data for Newfoundland and Labrador and New Brunswick includes public drug claims, and private insurers are first payers. The data linkage across LTC and drug data included all LTC residents with HD in New Brunswick and 88% of residents in Newfoundland and Labrador, with many drug claims; results of drug utilization analyses were therefore assumed to be representative.

In Project analysis 5, the LTC comparison population excluded Saskatchewan because drug claims for LTC residents cannot be identified in NPDUIS without data linkage.

Appendix C: Classification systems used in analyses

ATC: Anatomical Therapeutic Chemical.

ICD-10-CA: International Statistical Classification of Diseases and Related Health Problems, 10th Revision, Canada.

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