

CANADIAN PAEDIATRIC SURVEILLANCE PROGRAM

2018 Results



CPSP
CANADIAN PAEDIATRIC
SURVEILLANCE PROGRAM

Canadian
Paediatric
Society



Société
canadienne
de pédiatrie



Public Health
Agency of Canada

Agence de la santé
publique du Canada



Mission

To contribute to the improvement of the health of children and youth in Canada by national surveillance and research into childhood disorders that are high in disability, morbidity, mortality, and economic costs to society, despite their low frequency.

Canadian Paediatric Surveillance Program Annual Results

Surveillance is integral to the practice of public health. Public health surveillance, as defined by the World Health Organization, includes the systematic collection, collation, and analysis of data coupled with the timely dissemination of information for assessment and public health response. Integral to its public health mandate, the Canadian Paediatric Surveillance Program (CPSP) is committed to sharing valuable information obtained through its active surveillance of rare diseases and uncommon conditions in Canadian children and youth. Key results of CPSP multi-year studies and one-time surveys are published in this annual, bilingual report. These results highlight important findings and inform health professionals, researchers, and policy makers in developing strategies to improve the health of children and youth in Canada.

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Foreword

Federal Minister of Health

The Honourable Ginette Petitpas Taylor, P.C., M.P.



The Government of Canada is committed to supporting children's health. One way we demonstrate this commitment is through the Public Health Agency of Canada's collaboration with the Canadian Paediatric Society in supporting the Canadian Paediatric Surveillance Program.

For more than 20 years, the Canadian Paediatric Surveillance Program has collected data on rare and emerging childhood diseases and conditions as well as on public health issues affecting children. These data have informed countless advancements in the field of children's health.

This year's annual results report includes information on a study that is examining serious and life-threatening events associated with exposure to cannabis for non-medical purposes by children and youth (those under 18 years of age). Initiated shortly before the coming into force of the *Cannabis Act*, and scheduled to continue over the next two years, this study will contribute to the Government of Canada's efforts to better monitor the health and safety impacts of cannabis use. It will also help to raise awareness of the health and safety risks of cannabis.

I'd like to offer my sincere thanks to all the paediatricians and paediatric subspecialists who contribute to this annual collection of information. Through this collaboration, we've developed a solid and credible foundation of evidence for medical practitioners, researchers and policy makers to use in their work. Together, we can help improve medical care for children across the country.

Chief Public Health Officer of Canada

Dr. Theresa Tam



Researchers and health professionals across Canada have long relied on the valuable data generated through the Canadian Paediatric Surveillance Program (CPSP) to provide insight into rare disease risk factors, prevention practices, impacts of disease on children and youth, and how well treatments are working. This information is absolutely essential to the development and implementation of public health practices and policies.

Each year, the results presented in this report drive research and innovation in Canada and around the world, and encourage awareness and education, both within the medical community and the general public, about less-common paediatric disorders.

The CPSP is now in its third decade and currently covers a variety of conditions under study, contributing to ongoing improvements in child health across Canada.

As the Chief Public Health Officer of Canada, I am happy to present this annual report that so many of us rely on for up-to-date information on issues facing young Canadians today.

President of the Canadian Paediatric Society

Dr. Catherine Farrell

As a paediatric intensivist working at CHU Sainte-Justine for the past 29 years, my career has certainly brought me into close contact with families and young patients battling debilitating, life-threatening, and rare diseases. Diagnosing and treating rare diseases is very challenging when you consider the limited information and knowledge available to provide guidance to health care providers and reassurance to families.

As President of the Canadian Paediatric Society, I strongly believe in the value and importance of the CPSP which sheds light on rare conditions and emerging public health concerns. For example, the study on congenital Zika syndrome (page 19) helps to reassure Canadians that, although many people travel abroad every year, fewer than five cases of congenital Zika syndrome were reported over the duration of the study. The study on medically serious self-harm leading to ICU admissions (page 26) points to the significant burden associated with mental health conditions in Canadian children and youth and should inspire all of us to be engaged in early detection, effective intervention, and prevention strategies.

Another significant value of CPSP surveillance is providing clear evidence when a condition that was once thought to be rare is sadly no longer uncommon. The study on non-type 1 diabetes mellitus (page 21) obliges us, as a paediatric community, to focus on ways to reverse this concerning trend.

Information generated from CPSP studies and surveys provides evidence to determine the best path forward for our young patients and ensure they receive the best possible care. On behalf of the Canadian Paediatric Society and its Board of Directors, I would like to sincerely thank CPSP participants and investigators for all their contributions to this important network.



CPSP Chair

Dr. Jonathon Maguire

The CPSP had another banner year in 2018 with the implementation of three new multi-year studies and three one-time surveys. The CPSP continues to react in a timely fashion to new child and youth health threats and public health policies. A notable example is a new study on serious and life-threatening events from recreational cannabis which was launched one month prior to the legalization of cannabis in Canada (principal investigators: Drs. Richard Bélanger and Christina Grant). While legalization was directed at adults and regulations for edible cannabis products have not yet been approved, preliminary study results suggest that accidental consumption of edible cannabis products by children is resulting in serious injury requiring emergency care and hospitalization (page 32).

Another timely example is a new study which will compare outcomes from jurisdictional differences in policies for ophthalmia neonatorum prevention (principal investigators: Drs. Andrée-Anne Boisvert and Jesse Papenburg). In 2015, the Canadian Paediatric Society recommended against mandatory ocular prophylaxis with improved screening of pregnant women. Paediatricians have voiced concern that screening without ocular prophylaxis may result in an increase in the number of babies with ophthalmia neonatorum. This study is sure to contribute important, timely information to this ongoing public health debate (page 28).

We are fortunate in Canada to have public health surveillance systems like the CPSP which monitor health status, detect emerging health threats, and evaluate policies to improve the health of Canadians. The CPSP is the only national public health surveillance program for children and youth in Canada. It is particularly important for rare childhood diseases and rare complications of more common conditions where the collection of data from across Canada is necessary to obtain sufficient information to draw meaningful conclusions.

I would like to thank CPSP's Medical Affairs Director, Dr. Charlotte Moore Hepburn, and members of the Scientific Steering Committee who volunteer their time to ensure the highest scientific rigor for all CPSP studies. The CPSP relies entirely on the dedication of over 2,800 paediatricians and other health care providers who report new cases each month. On behalf of Canadian children and youth, I thank paediatricians across Canada who volunteer their time and expertise to report cases to the CPSP year after year. It would not happen without you!



Acknowledgements

The key strength of the Canadian Paediatric Surveillance Program is its commitment to improve the health of children and youth in Canada and around the world. This focus would not be possible without the participation of Canadian paediatricians, subspecialists, and other health care providers in the monthly collection of information on rare paediatric conditions, the principal investigators who design studies and analyse the data to provide knowledge and educational solutions, or the guidance of the Steering Committee members. We thank them all.

We also thank IMPACT (Immunization Monitoring Program ACTive) centres for their role in verifying the acute flaccid paralysis study data and for their support of the CPSP.

The strong partnership between the Canadian Paediatric Society and the Public Health Agency of Canada allows the program to grow in Canada and to take a leadership role on the international scene.

Funding

Funding for the CPSP is required to support program management. The surveillance program is funded through a combination of government support and unrestricted grants from Canadian charities, research institutions, hospitals, and corporations. All funding is provided to maintain and expand the program.

We gratefully acknowledge the financial support received in 2018 from the Public Health Agency of Canada's Centre for Surveillance and Applied Research, Health Canada's Marketed Health Products Directorate, and the following non-governmental sources:

- Genzyme
- The Chronic Pain Network, a Canadian Institutes for Health Research initiative

CPSP Steering Committee

Jonathon Maguire, MD (Chair)	Canadian Paediatric Society
Christine Armour, MD	Canadian College of Medical Geneticists (Liaison)
Peter Buck, DVM, MSc	Centre for Food-borne, Environmental and Zoonotic Infectious Diseases, Public Health Agency of Canada
Marie Adèle Davis, MBA	Canadian Paediatric Society
Elizabeth Donner, MD	Canadian Association of Child Neurology (Liaison)
Ciarán Duffy, MB	Paediatric Chairs of Canada (Liaison)
Joanne Embree, MD	IMPACT (Immunization Monitoring Program ACTive) (Liaison)
Krista Jangaard, MD	Canadian Paediatric Society
Carsten Kreuger, MD	Canadian Paediatric Society (Resident Representative)
Melanie Laffin Thibodeau, BCom	Canadian Paediatric Society
Sarah McDermott, MSc	Centre for Communicable Diseases and Infection Control, Public Health Agency of Canada
Charlotte Moore Hepburn, MD	Canadian Paediatric Society
Paul Muirhead, LL.M.	Consultant
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Jorge Pinzon, MD	Canadian Paediatric Society
Anne Rowan-Legg, MD	Canadian Paediatric Society
Miriam Santschi, MD	Canadian Paediatric Society
Jill Starkes, MD	Canadian Paediatric Society

The CPSP Scientific Steering Committee would like to extend a sincere thank you to Dr. Christine Armour who served a four-year term on the Committee as a representative for the Canadian College of Medical Geneticists. Her dedication and expertise on the Committee will be greatly missed and we wish her all the best in her future endeavours.



About the Canadian Paediatric Surveillance Program



Overview

The Canadian Paediatric Surveillance Program is a joint project of the Public Health Agency of Canada and the Canadian Paediatric Society that contributes to the improvement of the health of children and youth in Canada by national surveillance and research into childhood disorders that are high in disability, morbidity, and economic costs to society, despite their low frequency. The CPSP gathers data from over 2,800 paediatricians and paediatric subspecialists each month to monitor rare diseases and conditions in Canadian children.

Objectives

- Maintain an active national surveillance system that monitors low-frequency, high-impact conditions and diseases in Canadian children and youth
- Involve paediatricians, paediatric subspecialists, and other medical professionals in related disciplines in the surveillance of rare conditions that are of public health and medical importance
- Generate new knowledge into rare childhood disorders to facilitate improvements in treatment, prevention, and health-care planning
- Respond rapidly to public health emergencies relevant to Canadian children and youth by initiating rapid one-time surveys and new studies
- Participate in international paediatric surveillance efforts through the International Network of Paediatric Surveillance Units (INOPSU)

Surveillance

- The full surveillance process is summarized in Figure 1 and includes the 3Ds of surveillance: detection, deduction, and dissemination.
- Health surveillance can be defined as: the tracking of any health event or health determinant through the continuous collection of high-quality data (detection); the integration, analysis, and interpretation of the data (deduction) into surveillance products; and the dissemination of those surveillance products to those who need to know (dissemination).

Process

- Study teams from across Canada are encouraged to submit proposals for new studies or one-time surveys that meet the “criteria for submission,” available on the CPSP website at www.cpsp.cps.ca/apply-proposez.
- The CPSP Steering Committee then reviews the proposals on a biannual basis and selects those of highest medical and public health importance. Proposals are evaluated against set criteria and are subject

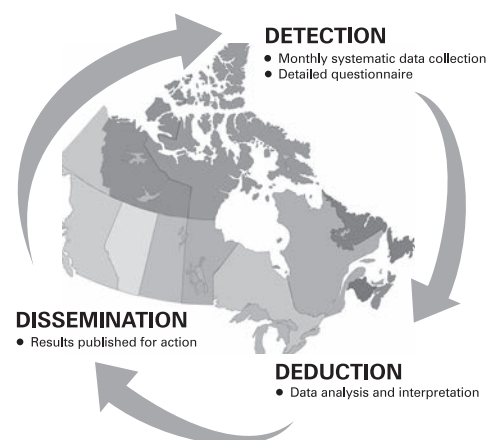
CPSP Quick Facts

Did you know?

- The CPSP celebrated its 22nd anniversary in 2018.
- The CPSP is comprised of over 2,800 dedicated paediatricians and paediatric subspecialists.
- Since its inception, the CPSP has studied 71 rare conditions/diseases and initiated 50 one-time surveys.
- Over 65 peer-reviewed manuscripts on study/survey results have been published in high-impact journals.
- The average monthly response rate is 80%.
- The average detailed questionnaire response rate varies between 80 to 90%.
- By December 2018, 82% of participants committed to receiving their monthly forms electronically.

Figure 1 – Surveillance process summary

Pan-Canadian health surveillance



to comprehensive feedback from the multidisciplinary Steering Committee, composed of representatives from the Public Health Agency of Canada, the Canadian Paediatric Society, former CPSP investigators, academic clinicians from diverse specialties, and community paediatricians.

- Each month, CPSP participants from across Canada receive a form listing the current conditions under study. Participants notify the program if they have seen any cases that meet the case definitions or have “nothing to report.” Participants are encouraged to report all cases, including suspect or probable cases. This sometimes leads to duplicate reporting but avoids missed cases.
- Participants who have seen a case are sent a detailed clinical questionnaire to complete and return to the CPSP.
- Once the detailed questionnaire is returned to the CPSP, it is stripped of all unique identifiers and sent to the investigators for data analysis. All notifications of potential cases are assessed against the case definition. Duplicates or cases that don’t meet the case definition are excluded.
- It is important to note that CPSP studies use anonymized data from patient charts; the study investigators have no direct contact with individual patients.
- The study team is responsible for data analysis, and for ensuring that a solid knowledge translation plan is in place to disseminate the results in a timely and effective manner.
- Study results are published annually and acted upon to improve the health of Canadian children and youth. For example, CPSP study results help to warn of emergent public health issues, identify safety hazards, mobilize knowledge on rare diseases/conditions, and inform new policies and guidelines.

Response rates

The CPSP’s average national monthly response rate is 80% and the average detailed questionnaire completion rate varies between 80 to 90%.

TABLE 1 – Initial response rates (%) and number of participants for 2018

Provinces/territories	Reporting rates (%) [*]	Number of participants [†]
Alberta (AB)	76	387
British Columbia (BC)	74	288
Manitoba (MB)	80	111
New Brunswick (NB)	91	28
Newfoundland and Labrador (NL)	82	48
Northwest Territories (NT)	97	3
Nova Scotia (NS)	87	87
Nunavut (NU)	25	3
Ontario (ON)	77	1,025
Prince Edward Island (PE)	92	9
Quebec (QC)	78	569
Saskatchewan (SK)	76	63
Yukon (YT)	100	1
Canada	78	2,622

^{*} The CPSP national monthly reporting rate averages 80%. Every effort is made to maximize reporting, and annual response rates are subject to change due to delays in reporting.

[†] The total number of individual CPSP participants is over 2,800. However, in this table, the number of CPSP participants in Canada is calculated based on both individual and group reporting. When a group designate responds to the CPSP on behalf of group members, it is counted as one response.

TABLE 2 – National initial response rates 2014–2018

Reporting year	Reporting rates (%)
2014	81
2015	82
2016	79
2017	82
2018	78

TABLE 3 – 2018 detailed questionnaire completion rates as of June 13, 2019

Studies/conditions	Notifications of potential cases*	Pending	% Completion rate
Acute flaccid paralysis	91	12	87
Adverse drug reactions – serious and life-threatening	23	4	83
Complex regional pain syndrome in Canadian children and youth	130	24	82
Congenital Zika syndrome in infants in Canada	<5	<5	—
Incidence trends of type 2 diabetes, medication-induced diabetes, and monogenic diabetes in Canadian children	277	59	79
Infantile and later-onset paediatric Pompe disease (glycogen storage disease type II)	13	1	92
Medically serious self-harm in youth requiring ICU admission	60	9	85
Ophthalmia neonatorum caused by <i>N gonorrhoeae</i> or <i>C trachomatis</i>	<5	<5	—
Rh sensitization	17	2	88
Serious and life-threatening events associated with non-medical (recreational) cannabis use in Canadian children and youth	13	1	92
Severe microcephaly	10	4	60
Severe obesity and global developmental delay in preschool children	36	9	75
Total number of cases (all studies)	670	125	83

* Excludes case notifications from Quebec from August 1 to December 31, 2018

Glossary of terms in study results

Reported: Notifications of potential cases received by the CPSP

Reported from Quebec August to December: In mid-2018, the CPSP became aware of a change in Quebec legislation that affected the ability of the program to collect detailed information from physicians who practise in that province. An interim measure was implemented, asking Quebec participants to defer returning their detailed questionnaires until a new process is defined. Cases notified by Quebec physicians prior to August 1, 2018 are eligible to be included in the data analysis.

Duplicates: Cases reported by more than one participant

Excluded: Cases not meeting the case definition

Pending: Detailed reports not received or not yet verified as meeting the case definition

Met case definition: Cases verified as meeting the case definition, not including reports from Quebec from August to December 2018, duplicates, excluded, and pending cases

International Network of Paediatric Surveillance Units

The CPSP offers an opportunity for international collaboration with other paediatric surveillance units worldwide, through the International Network of Paediatric Surveillance Units (INOPSU). The network provides a successful and easily accessible platform for international surveillance. No other network enables international comparisons of demographics, diagnosis, treatments, and outcomes for rare childhood conditions.

Established in 1998, INOPSU now includes 16 paediatric surveillance units among its membership. Full member countries are Australia, Canada, Netherlands, New Zealand, Switzerland, United Kingdom, and Wales. Affiliate members are: Germany, Belgium, Scotland, Portugal, Ireland, and Greece/Cyprus as well as the British Ophthalmology Surveillance Unit, the British Neurology Surveillance Unit, and the UK Obstetrics Surveillance System.



Many of the paediatric surveillance units have been collecting data on rare childhood conditions for 20 years or more. Over 300 rare conditions have been studied to date, including rare infectious and vaccine-preventable diseases, mental health disorders, child injuries, and immunological conditions. The network encompasses approximately 10,000 child health care providers who voluntarily contribute data on these rare diseases every month.

INOPSU 10th scientific conference

On March 13, 2018 INOPSU held its 10th scientific conference in Glasgow, Scotland. The conference explored paediatric rare disease surveillance and its global impact on public health and clinical practice. Over 150 delegates attended to hear scientific data presented by world-leading clinicians, scientists, and policy makers. Presentations considered a range of topics such as the central role paediatric surveillance has played in the identification of emerging infectious diseases, the importance of international collaboration on understanding the natural history of rare diseases, and the continued need for monitoring of emerging and re-emerging diseases such as *E. coli* O157.

CPSP-related presentations included the following:

- Listeriosis in neonates and infants in Switzerland and Canada by Ms. Mirjam Mäusezahl-Feuz, INOPSU Convener, Switzerland
- Monitoring for hypoglycemic newborns – Should we expand our risk categories? by Dr. Michael Flavin, Queen's University, Canada
- Medical assistance in dying: A Canadian perspective by Dr. Charlotte Moore Hepburn, Director of Medical Affairs, Canadian Paediatric Society, Canada

In memory of the late Dr. Danielle Grenier, Director of Medical Affairs of the Canadian Paediatric Society, the Danielle Grenier Prize is awarded during the INOPSU scientific session to recognize excellence in paediatric surveillance research and knowledge translation. The CPSP congratulates this year's winner, Dr. Ifeanyichukwu Okike from the Derbyshire Children's Hospital in the United Kingdom who presented: Neonatal meningitis – Developing guidelines from surveillance data.

More information on INOPSU can be found at <http://www.inopsu.com/>.



Dr. Ifeanyichukwu Okike

Surveillance Studies in 2018

Acute flaccid paralysis

Ongoing study since January 1996



Catherine Dickson

Principal investigator

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? Question

Did Canada maintain its polio-free status in 2018?

! Importance

- Acute flaccid paralysis (AFP) surveillance is the cornerstone of monitoring for polio, in light of ongoing transmission of wild poliovirus in a few countries around the world.
- Canada conducts AFP surveillance in children under 15 years of age, in accordance with World Health Organization (WHO) recommendations and standards of practice.

↻ Methodology

The complete protocol can be accessed at www.cpsp.cps.ca/surveillance.

Case definition

Acute onset of focal weakness or paralysis characterized as flaccid (reduced tone) without other obvious cause (e.g., trauma) in a child less than 15 years of age. Transient weakness (e.g., post-ictal weakness) does not meet the case definition.

Unique to this study

Cases are captured through both the Canadian Paediatric Surveillance Program (CPSP) and Canada's Immunization Monitoring Program ACTIVE (IMPACT) based in 12 tertiary care paediatric centres. Given the concerns surrounding increased detection of acute flaccid myelitis (AFM) in the United States in the fall of 2018, the CPSP heightened awareness among providers and hospitals to increase vigilance in reporting.

✓ Results – January to December 2018

Note: Due to reporting delay, this report represents a snapshot as of April 11, 2019 for cases that occurred between January 1, 2018 and December 31, 2018.

TABLE 1 – AFP cases in 2018

Reported	Reported to CPSP* from Quebec August to December	Duplicates	Excluded	Pending	Met case definition
100	9	13	5	24	49

* Does not include cases reported through IMPACT centres in Quebec which have approvals to send AFP detailed questionnaires to the CPSP. Therefore, AFP cases reported by Quebec IMPACT centres are eligible for data analysis in this report.

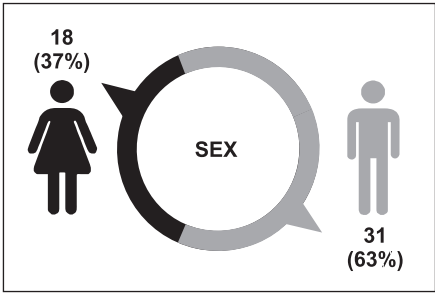
Cases that met the case definition

- In total, 100 reports of sudden onset muscle weakness in children were provided to the Public Health Agency of Canada: 42 (42%) cases were reported through the CPSP network and 58 (58%) cases were reported through IMPACT.

- All AFP cases were adjudicated against the national AFP and polio case definitions.
- At the time of analysis, 49 cases were verified as meeting the AFP case definition in 2018; none were assessed to be polio.
- The average time from case onset to reporting was 87 days (range: 10 to 298).

Demographics

- Patient sex was male in 31 (63%) cases and female in 18 (37%) cases.
- Cases ranged in age from younger than 1 year to 14 years, with a mean of 5.7 years (95% CI 4.6–6.8) and a median of 4.7 years.



Presentation

- Hospitalization: All 49 (100%) cases were hospitalized and length of stay ranged from 2 to 70 days with a mean of 15 days (95% CI 11.1–19.4) and a median of 11 days.
- Vaccinations: 37 (76%) cases were up-to-date for their polio vaccinations while 6 (12%) cases were not. The remaining 6 (12%) cases had unknown polio vaccination information.
- Diagnoses: All types of AFP are monitored in Canada, including AFM for which an increased number of cases were reported in the United States in the Fall of 2018. The following diagnoses were reported by physicians as types of AFP: 15 (31%) cases were Guillain-Barré syndrome and 13 (27%) were transverse myelitis. Diagnoses for the remaining 21 (43%) cases included: acute disseminated encephalomyelitis, AFM, acute extensive myelitis or flaccid paralysis, chronic inflammatory demyelinating polyneuropathy, Epstein-Barr virus, myeloradiculopathy, infant botulism, and right brachial plexopathy.
- No stool samples were positive for polio.

Treatment and outcomes

- Of the 40 (82%) cases that had outcome documented at the time of initial report, 32 (80%) had partially recovered with residual weakness, 5 (13%) had not recovered, and the rest had fully recovered.
- Eighteen (37%) cases had the clinical outcome reported at least 60 days after the onset of paralysis or weakness; 13 (72%) had partially recovered and the remaining 5 cases had either fully recovered or had not recovered.

TABLE 2 – Measure of Canada’s performance against WHO AFP surveillance performance indicators in 2018¹

Number of cases	Incidence rate [*]	% with adequate stool sample ^{‡†}	% with 60-day follow-up [‡]
49	0.76	35%	37%

^{*} Target is 1.0 AFP case per 100,000 population in those less than 15 years of age

[†] Target is at least 80% of cases have adequate stool sampling

[‡] Target is at least 80% have follow-up examination for residual paralysis at least 60 days after onset

Study limitations

- As with any voluntary reporting surveillance system, the CPSP recognizes that reporting on minimum incidence rates can have limitations, including under-representation of the disease/condition in the population. It is possible that some groups of children were missed, for example those who live in rural or remote areas, as they may be less likely to receive timely specialist care.
- Case-level surveillance data are extracted from patient charts following the clinical encounter. Data elements, such as pending test results or travel and immunization history, may not have been available at the point of care and therefore may be absent from the surveillance totals. However, surveillance serves an important purpose and provides rich clinical data that allow Canada to maintain its polio-free status.
- Stool samples in patients with AFP are sometimes difficult to obtain due to the nature of the patient’s symptoms, including constipation. Additionally, rapid availability of advanced diagnostic testing often identifies the diagnosis prior to the collection of the stool sample.

Conclusions

There was sufficient evidence to suggest that no polio cases occurred in Canada even though Canada did not meet the WHO performance indicators for national AFP surveillance in 2018.

Anticipated study impact

- Canada’s polio-free status remains intact.
- In its efforts to continue to support the Polio Endgame Strategy and strengthen AFP surveillance, Canada is updating the national case definition for polio.

Acknowledgements

The investigators would like to thank everyone who participated in collecting the data. They would also like to acknowledge the excellent work of Noémie Desmarceaux, Lauren Clow, and Diane MacDonald.

1. Detailed information on WHO surveillance performance indicators can be found at <http://polioeradication.org/polio-today/polio-now/surveillance-indicators/>
 2. Adequate stool sample refers to one stool sample taken within 14 days of paralysis onset.

Adverse drug reactions – serious and life-threatening

Ongoing study since January 2004



Sally Pepper

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? Question

What serious and life-threatening events suspected to be related to adverse drug reactions (ADRs) in children and youth were reported in 2018?

! Importance

- Only a minority of prescribed pharmaceuticals on the market in North America have been tested in paediatric patients, and most of them are used without the benefit of adequate and/or specific guidance on safety or efficacy in this population.
- Post-marketing surveillance is essential for detection of ADRs, and contributes to the ongoing monitoring of the benefit-risk profile of health products used in children.

➡ Methodology

The complete protocol can be accessed at www.cpsp.cps.ca/surveillance.

Case definition

Serious and life-threatening adverse drug reactions* in an infant or child up to the age of 18 years, associated with the use of prescription, non-prescription, biological (immunoglobulin) products, complementary medicines (including herbals), and radiopharmaceutical products.

* Noxious and unintended severe response to a drug, which occurs at any dose and results in emergency observation, hospitalization, persistent or significant disability, or death

Exclusion criteria

Reactions to medical devices, blood products, (platelets, red cells and single-donor plasma), vaccines, poisonings or self-administered overdoses

Unique to this study

Significant results for the ADR study contribute to the monthly ADR Tips distributed by the Canadian Paediatric Surveillance Program (CPSP).

✓ Results – January to December 2018

TABLE 1 – ADR cases in 2018					
Reported	Reported from Quebec August to December	Duplicates	Excluded	Pending	Met case definition
22	0	0	2	0	20

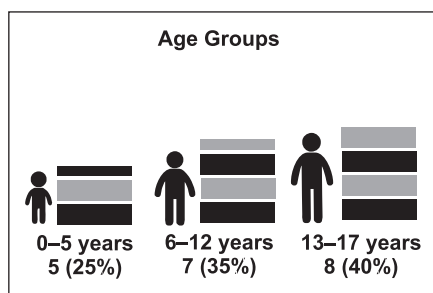
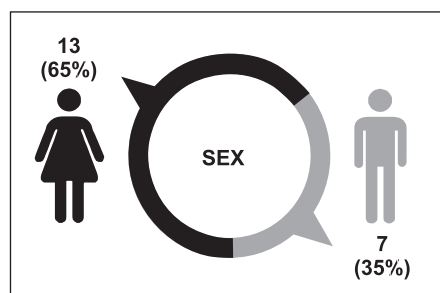
Cases that met the case definition

- At the time of analysis, 20 suspected serious and/or life-threatening paediatric ADR cases were verified as meeting the case definition in 2018.
- In fewer than five reports, more than one product was suspected of causing the adverse reaction.

- Classes of health products, as outlined by the Anatomical Therapeutic Chemical (ATC) classification system, most frequently suspected of causing the adverse reaction(s) were antibacterials, immunosuppressants, and psycholeptics.
- Anaesthetics, analgesics, anti-inflammatory and antirheumatic products, natural health products, pituitary and hypothalamic hormones and analogues, psychoanaleptics, and other therapeutic agents were each involved in fewer than five case reports.

Demographics

- Patient sex was male in 7 (35%) cases and female in 13 (65%) cases.
- Age ranges of cases were as follows: 5 (25%) aged 0 to 5 years, 7 (35%) aged 6 to 12 years, and 8 (40%) aged 13 to 17 years.



Presentation

- The 20 cases that met the case definition were classified as serious according to the following criteria (more than one cause for classification was provided in 11 reports): 9 (45%) cases were considered to be life-threatening and 15 (75%) cases required hospitalization.
- The majority of the adverse reaction reports described nervous system disorders. Reactions reported under this category include seizure, decreased level of consciousness, extrapyramidal disorder, and neuroleptic malignant syndrome, among others. This finding represents a new outcome, as the majority of the reactions reported through the CPSP since 2004 have been skin and subcutaneous tissue disorders.
- As in prior years, the majority of the reports describe reactions generally documented in the approved Canadian product monograph (CPM) or other drug information references.

Treatment and outcomes

- The outcome was known in most of the 20 cases, with the majority of patients (75%, 15/20) experiencing a full recovery.
- Fewer than five cases reported disability and there were fewer than five deaths.

Study limitations

- All adverse reactions to health products are considered suspicions as a definite causal association often cannot be determined. The true incidence of adverse reactions is unknown because they remain under-reported and total patient exposure is unknown.
- As with any voluntary reporting surveillance system, the CPSP recognizes that reporting on minimum incidence rates can have limitations, including under-representation of adverse reactions in the population. It is possible that some groups of children were missed, for example those who live in rural or remote areas, as they may be less likely to receive timely specialist care.
- Case-level surveillance data are extracted from patient charts following the clinical encounter. Data elements, such as laboratory investigations, pre-existing medical conditions, and relevant components of the diagnostic assessment, may not have been available at the point of care and therefore may be absent from the surveillance totals. However, surveillance serves an important purpose and provides rich clinical data that will allow for a better understanding of serious and life-threatening paediatric ADRs.

TABLE 2 – Suspect health products in 2018	
Class of health product	Name of health product
Anaesthetics	Propofol
Analgesics	Hydromorphone
Antibacterials	Ceftriaxone, co-trimoxazole*
Antiepileptics	Lamotrigine
Anti-inflammatory and antirheumatic products	Ibuprofen
Antimycobacterials	Dapsone
Antineoplastic agents	Cytarabine, imatinib
Immunosuppressants	Abatacept, anakinra, methotrexate
Natural health products	Mixture of calming oils (arnica, clove, evening primrose, German chamomile, grapeseed, lavender, orange, peppermint)
Other therapeutic agents	Deferasirox, naloxone
Pituitary & hypothalamic hormones & analogues	Desmopressin, somatropin
Psychoanaleptics	Fluoxetine, methylphenidate
Psycholeptics	Quetiapine, risperidone, zuclopenthixol

* Combination product containing two or more active ingredients

Conclusions

- Classes of health products most frequently suspected of causing adverse reaction(s) reported in 2018 were antibacterials, immunosuppressants, and psycholeptics.
- Since the implementation of the CPSP surveillance for adverse reactions in 2004, the product classes most frequently associated with suspect products have been antibacterials for systemic use, antiepileptics, and psychoanaleptics. The most frequently reported suspect drugs in these classes are amoxicillin, carbamazepine, and methylphenidate respectively. No reports meeting the study criteria were received in 2018 for amoxicillin and carbamazepine.

Anticipated study impact

- Health Canada recognizes the need to strengthen information related to paediatric health, as the use of medications to treat children is increasing, and the safety and efficacy of these medications may be significantly different in paediatric patients than in adult patients.^{1,2} The ongoing sharing of safety information through voluntary reporting of ADRs from various sources such as the CPSP is valuable to Health Canada as it contributes to ongoing monitoring of the benefit-risk profile of health products used in children and can thus result in the implementation of risk mitigation measures.
- In acknowledgement of the importance of safety information provided by ADR reporting, Health Canada is implementing Vanessa's Law, an amendment to the *Food & Drugs Act* that requires certain health care institutions to identify and report serious adverse drug reactions to the federal regulator. A key objective of mandatory reporting is to improve the quality and quantity of serious ADR reports, and to expand on the real-world data available to monitor the safety of health products used in children.

Acknowledgements

The assistance of Lynn Macdonald is greatly appreciated.

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2. Abi Khaled L, Ahmad F, Brogan T, et al. Prescription medicine use by one million Canadian children. *Paediatr Child Health* 2003;8(A):6A-56A

Complex regional pain syndrome in Canadian children and youth

September 2017 to August 2019



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? Question

- What are the minimum incidence and geographic distribution of complex regional pain syndrome (CRPS) in the Canadian paediatric population?
- What are the pathways of referral, clinical presentation, diagnostic interventions, and recommended interventions by paediatricians and pain specialists?

! Importance

- CRPS is a rare chronic severe pain condition that involves peripheral, central, and autonomic nervous system and immune system mechanisms. It results in significant functional impairment and debilitating symptoms. The persistent and severe pain results in psychological, physical, and neurological structural and functional changes.
- Few interventions for CRPS have been formally evaluated in the paediatric population.

➡ Methodology

The complete protocol can be accessed at www.cpsp.cps.ca/surveillance.

Case definition

A patient presenting between the ages of 2 and 18 years (up to the 18th birthday) with a new diagnosis of CRPS, meeting the following International Association for the Study of Pain clinical diagnostic criteria:

1. Continuing pain, which is disproportionate to any inciting event
2. Reports at least one symptom in at least three of the following four categories:
 - **Sensory:** hyperesthesia and/or allodynia
 - **Vasomotor:** temperature asymmetry and/or skin color changes and/or skin color asymmetry
 - **Sudomotor/Edema:** edema and/or sweating changes and/or sweating asymmetry
 - **Motor/Trophic:** decreased range of motion and/or motor dysfunction (weakness, tremor, dystonia) and/or trophic changes (hair, nail, skin)
3. Displays at least one sign at time of evaluation in at least two of the following four categories:
 - **Sensory:** hyperalgesia (to pinprick) and/or allodynia (to light touch and/or temperature sensation and/or deep somatic pressure and/or joint movement)
 - **Vasomotor:** temperature asymmetry ($>1^{\circ}\text{C}$) and/or skin color changes and/or asymmetry
 - **Sudomotor/Edema:** edema and/or sweating changes and/or sweating asymmetry
 - **Motor/Trophic:** decreased range of motion and/or motor dysfunction (weakness, tremor, dystonia) and/or trophic changes (hair, nail, skin)

Exclusion criteria

Presence of another diagnosis that better explains the signs and symptoms



Results – January to December 2018

TABLE 1 – CRPS cases in 2018

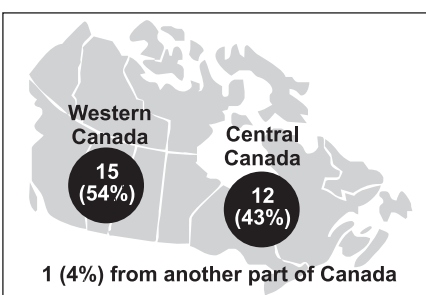
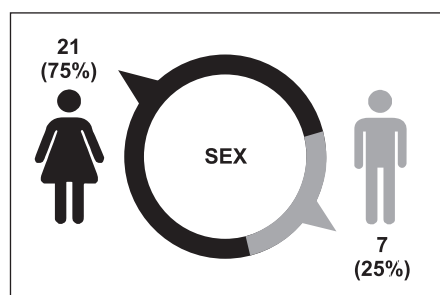
Reported	Reported from Quebec August to December	Duplicates	Excluded	Pending	Met case definition
73	2	0	12	31	28

Cases that met the case definition

- At the time of analysis, 28 cases of CRPS were verified as meeting the case definition in 2018.

Demographics

- Patient sex was female in 21 (75%) cases and male in 7 (25%) cases.
- The mean age of cases was 11.7 years (SD 2.6).
- The geographic distribution of cases was: 15 (54%) from Western Canada, 12 (43%) from Central Canada, and the remaining case was from another part of Canada.
- Population groups were as follows: 24 (86%) were White and the remaining cases were from other population groups.



Presentation

- The average number of months between onset and diagnosis of CRPS was 2.1 months (SD 2.5).
- The average body mass index was 24.9 (SD 4.4).
- Average pain intensity over the past week was reported as severe in 20 (71%) cases and moderate in 6 (21%) cases.
- The most common inciting/triggering event for CRPS was trauma/injury in 16 (57%) cases. There was no known trigger in 8 (29%) cases.
- CRPS presented more commonly in the lower limbs (20, 72%) and affected both sides of the body equally.
- Patients reported symptoms in the following categories: 25 (89%) motor/trophic, 25 (89%) sensory, 21 (75%) sudomotor/edema, and 20 (71%) vasomotor.
- At the time of evaluation, patients displayed the following signs: 25 (89%) motor/trophic, 23 (82%) sensory, 18 (64%) vasomotor, and 17 (61%) sudomotor/edema.
- The majority of cases missed school (16, 57%).
- Symptoms of CRPS had a functional impact on patients in the following areas: 28 (100%) physical activity, 17 (61%) sleep, 12 (43%) social activities, 11 (39%) mood, 10 (36%) family function, 9 (32%) high-level sport, and 7 (25%) school achievement.

Treatment and outcomes

- Most CRPS patients received pain medications and adjuvants, including the following: 24/28 (86%) NSAIDs, 18/27 (67%) acetaminophen, 11/27 (41%) gabapentinoids, 8/25 (32%) topicals, and 5/25 (20%) tricyclic antidepressants.
- The most common complementary therapy reported is chiropractic (5/28, 18%).
- CRPS patients received the following treatments: 17 (61%) pain education, 15 (54%) psychological strategies, 13 (46%) fitness/exercise, 13 (46%) desensitization, 9 (32%) bracing/ankle-foot orthosis/boot, 6 (21%) TENS machine, and 5 (18%) graded motor imagery.
- Nutritional supplements were received in 5 (18%) cases.
- CRPS patients were referred to the following health care providers: general paediatricians, physiotherapists, psychologists, occupational therapists, psychiatrists, orthopedic specialists, neurologists, and multidisciplinary paediatric pain clinics (Note: a paediatric pain clinic commonly involves a paediatrician or anaesthesiologist, nurse, physiotherapist, and psychologist).

Study limitations

- As with any voluntary reporting surveillance system, the CPSP recognizes that reporting on minimum incidence rates can have limitations, including under-representation of the disease/condition in the population. It is possible that some groups of children were missed, for example those who live in rural or remote areas, as they may be less likely to receive timely specialist care.

- Case-level surveillance data are extracted from patient charts following the clinical encounter. Data elements, such as social history or special investigations, may not have been available at the point of care and therefore may be absent from the surveillance totals. However, surveillance serves an important purpose and provides rich clinical data that will allow for a better understanding of the Canadian-specific epidemiology and the impact of CRPS.



Conclusions

- At the time of analysis, 28 cases of CRPS were verified as meeting the case definition in 2018.
- CRPS affected more females than males and presented more commonly in the lower limbs, consistent with the current CRPS adult literature.
- The majority of cases affected were White.
- The majority of CRPS signs and symptoms reported were sensory and motor/trophic.
- The average time between onset and diagnosis of CRPS was 2.1 months (SD 2.5).
- Treatment involved a combination of medications, complementary medicines, nutritional supplementation, and other therapies such as pain education and psychological strategies.



Anticipated study impact

- Study results will help determine the minimum incidence of CRPS and highlight current resource needs in Canada.
- The study will help identify patient demographics, triggers, or risk factors associated with CRPS.
- Results will be used to promote early recognition and treatment to benefit patient recovery.

Acknowledgements

Thanks to all the collaborating Canadian paediatric pain clinics, the CPSP, and CPSP participants. We gratefully acknowledge funding for this study from the Chronic Pain Network.

Congenital Zika syndrome in infants in Canada

March 2017 to February 2019



Shaun Morris

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? Question

What is the minimum incidence of infants born with congenital Zika syndrome (CZS) in Canada and what is the spectrum of clinical manifestations and abnormalities seen in these infants?

! Importance

- In October 2015, an increased incidence of microcephaly was noted in northeastern Brazil. Further investigations noted an increase in severe microcephaly and other neurological disorders among newborns born to mothers with Zika virus infection.
- While severe microcephaly was the first major congenital anomaly linked with Zika virus infection during pregnancy, a wide range of congenital anomalies have been described. As a result of the spectrum of clinical manifestations and abnormalities seen in infants born to Zika virus-infected mothers, the term congenital Zika syndrome has been developed. Importantly, some newborns born to mothers infected with Zika virus have neurological abnormalities with a normal head circumference.
- Surveillance for CZS in Canada through the Canadian Paediatric Surveillance Program (CPSP) started in March of 2017. This project is complementary to the CPSP surveillance for severe microcephaly.

↻ Methodology

The complete protocol can be accessed at www.cpsp.cps.ca/surveillance.

Case definition

An infant less than 12 months of age who presents with the following criteria:

- Microcephaly, defined as head circumference less than two standard deviations for gestational age and sex according to the standardized reference percentile*
OR
- Other congenital anomalies and malformations consistent with congenital Zika syndrome including malformations of the central nervous system, such as intracranial calcifications, structural brain or eye abnormalities, or other congenital central nervous system-related abnormalities (not explained by another etiology†)
AND
- A maternal history that includes an epidemiologic linkage‡ to Zika virus OR a positive or inconclusive Zika virus laboratory test
OR
- An infant with a positive or inconclusive Zika virus laboratory test

* If there is a case of severe microcephaly suspected to be associated with Zika virus then a questionnaire for the severe microcephaly study and the congenital Zika syndrome study should be completed (i.e., if the case meets both case definitions).

† Other etiologies that should be considered include other congenital infections such as syphilis, toxoplasmosis, rubella, cytomegalovirus, varicella zoster, parvovirus B19, and herpes simplex virus. An assessment of potential genetic and other teratogenic causes of the congenital anomalies should also be considered.

‡ Epidemiological linkage means: travelled to, or resided in, an area with active Zika virus transmission during her pregnancy; OR had unprotected sex during pregnancy with a partner who resided in, or traveled to, an area with active Zika virus transmission.

Unique to this study

A CPSP study examining the incidence and epidemiology of severe microcephaly in Canada took place from June 2016 to May 2018. For cases of severe microcephaly suspected to be associated with Zika virus, CPSP participants were asked to report using both the severe microcephaly questionnaire AND the CZS questionnaire. There was cross-representation of principal and co-investigators on the research teams to ensure that all cases were appropriately identified and analyzed.

Results – January to December 2018

TABLE 1 – CZS cases in 2018					
Reported	Reported from Quebec August to December	Duplicates	Excluded	Pending	Met case definition
<5	0	<5	<5	<5	<5

Cases that met the case definition

- Since data collection was initiated, fewer than five cases of CZS have been verified as meeting the case definition in Canada.

Demographics

- As per CPSP policy, case numbers and data for fewer than five cases cannot be presented.

Presentation, treatment, and outcomes

While specific information on this study cannot be presented at the current time due to the small number of cases, available literature demonstrates that CZS consists of:

- Severe microcephaly in which the skull has partially collapsed
- Decreased brain tissue with a specific pattern of brain damage, including subcortical calcifications
- Damage to the structures of the eye, including but not limited to macular scarring and focal pigmentary retinal mottling
- Congenital contractures
- Hypertonia

According to the Public Health Agency of Canada, as of August 31, 2018, 569 travel-related cases and 4 sexually transmitted cases of Zika virus infection have been reported in Canada since June 2015. Studies show that among completed pregnancies with laboratory evidence of infection with Zika virus, about 6% of fetuses or infants had evidence of Zika-associated birth defects. Among pregnant women with Zika virus infection in the first trimester, about 11% of fetuses or infants had evidence of Zika-associated birth defects.

Study limitations

- As with any voluntary reporting surveillance system, the CPSP recognizes that reporting on minimum incidence rates can have limitations, including under-representation of the disease/condition in the population. It is possible that some groups of children were missed, for example those who live in rural or remote areas, as they may be less likely to receive timely specialist care.
- Case-level surveillance data are extracted from patient charts following the clinical encounter. Data elements, for example pending test results, parental travel history, physical examination, and relevant components of the diagnostic assessment, may not have been available at the point of care and therefore may be absent from the surveillance totals. However, surveillance serves an important purpose and provides rich clinical data that will allow us to better understand the Canadian-specific epidemiology and impact of Zika virus.

Conclusions

CZS is rare in Canada. Thus far, fewer than five cases of CZS have been verified as meeting the case definition in this CPSP study.

Anticipated study impact

- While CZS has been rare thus far in Canada, this study will provide valuable clinical and epidemiological information on cases that may occur following maternal infection via travel, sexual transmission, or other modes.
- These data will supplement any other data collected via provincial or territorial reportable disease programs.

Acknowledgements

The investigators and the CPSP would like to thank all physicians who have reported cases to this study.

Incidence trends of type 2 diabetes, medication-induced diabetes, and monogenic diabetes in Canadian children

June 2017 to May 2019



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? Question

- What are the minimum incidence and the 10-year minimum incidence trends of non-type 1 diabetes mellitus (NT1DM) and its subtypes (type 2 diabetes [T2D], medication induced diabetes [MID], and monogenic diabetes) in Canada?
- What are the childhood-onset T2D risk factors, clinical characteristics, and diabetes-related complications, as well as differences in approaches to treatment across Canada?

! Importance

- The incidence of childhood-onset T2D is increasing. With Canadian data on T2D incidence in children and youth from 2006 to 2008, this second Canadian Paediatric Surveillance Program (CPSP) study will provide national incidence trend data over a 10-year period.
- This surveillance study will produce the first-ever 10-year incidence trends for other forms of NT1DM including MID and monogenic diabetes.

➡ Methodology

The complete protocol can be accessed at www.cpsp.cps.ca/surveillance.

Case definition

A new or revised* diagnosis of non-type 1 diabetes (NT1DM) in a patient less than 18 years of age with clinical features that are **not** consistent with classic type 1 diabetes (defined as a child with symptomatic acute hyperglycemia).

* A revised diagnosis occurs when a child previously diagnosed with type 1 diabetes mellitus receives a "revised" diagnosis of non-type 1 diabetes based on clinical progression and/or results of investigations.

Diabetes is defined based on the Diabetes Canada Guidelines:

- Fasting plasma glucose (FPG) ≥ 7.0 mmol/L[†] or
- Random plasma glucose ≥ 11.1 mmol/L[†] or
- Two-hour plasma glucose ≥ 11.1 mmol/L[†] after a standard oral glucose tolerance test

† Requires a second, confirmatory test if child is asymptomatic

Clinical features suggestive of non-type 1 diabetes mellitus are listed below:

- a) Obesity (body mass index >95 th percentile for age and gender)
- b) Family history of type 2 diabetes in a first- or second-degree relative(s)
- c) Belonging to a high-risk ethnic group (e.g., Indigenous, Black, Latin American, South-Asian)
- d) A history of exposure to diabetes *in utero* (diagnosed before or during pregnancy)
- e) Acanthosis nigricans
- f) Polycystic ovarian syndrome

- g) Diabetes in a person with a syndrome often associated with type 2 diabetes (Prader-Willi syndrome)
- h) Diabetes in a non-obese patient with at least one first-degree relative with diabetes
- i) Diabetes diagnosed in a neonate/infant less than 6 months of age
- j) Minimal or no insulin requirement with a normal or near normal A1c level (4–6%) one year after diagnosis
- k) A diagnosis of diabetes while on medical therapy with a known diabetogenic medication (e.g., glucocorticoids, L-asparaginase, cyclosporine, tacrolimus, atypical antipsychotic, anticonvulsant)

Exclusion criteria

Do not report patients with cystic fibrosis-related diabetes, pregnant teenagers with gestational diabetes, and patients in critical care settings requiring **short-term** insulin therapy for stress hyperglycemia.

Unique to this study

- To produce the most accurate incidence estimate of childhood-onset T2D, this study will also use data from the Canadian Primary Care Sentinel Surveillance Network (CPCSSN) to identify new cases of childhood-onset T2D seen by participating primary care providers (e.g., family physicians) that would not be reported by paediatricians or paediatric endocrinologists.
- Reporting physicians have the option of accessing free specialized pancreatic autoantibody testing accessed via the Barbara Davis Center for Childhood Diabetes (Denver, Colorado) if they feel that the additional testing would help with the classification of diabetes subtype, and they do not have access to this testing via their provincial laboratory services. This part of the study is not conducted through the CPSP and requires patient consent.

✓ Results – January to December 2018

TABLE 1 – NT1DM cases in 2018

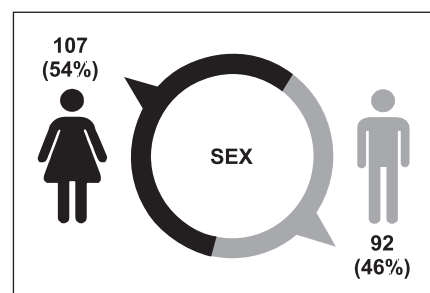
Reported	Reported from Quebec August to December	Duplicates	Excluded	Pending	Met case definition
266	11	0	9	47	199

Cases that met the case definition

- At the time of analysis, 199 cases were verified as meeting the case definition for NT1DM in 2018.

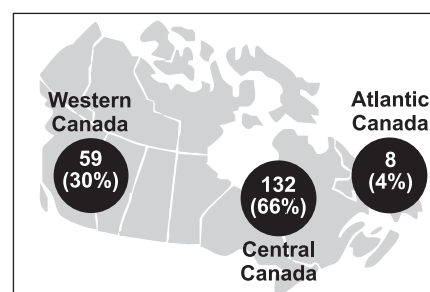
Demographics

- Patient sex was female in 107 (54%) cases and male in 92 (46%) cases.
- The geographic distribution of cases was: 59 (30%) from Western Canada, 132 (66%) from Central Canada, and 8 (4%) from Atlantic Canada.
- Population groups were indicated for 196 cases with the majority of cases being from the following groups: 76 (39%) Indigenous, 65 (33%) White, and 21 (11%) South Asian (Bangladeshi, Punjabi, Sri Lankan, Indian).



Presentation

- The reporting physician made a clinical diagnosis of diabetes subtype in 196 cases where: 140 (71%) were T2D, 34 (17%) were MID, and 7 (4%) were confirmed or suspected monogenic diabetes. There were 15 (8%) cases where the diagnosis of a diabetes subtype was unknown or unconfirmed, and 3 cases where data was missing.
- Patients were asymptomatic in 98/189 (52%) cases.
- Polyuria was present at diagnosis in 75/189 (40%) cases and polydipsia in 71/189 (38%) cases.
- Diabetic ketoacidosis (DKA) was described in 14/189 (7%) cases. Fewer than five cases each presented with a hyperglycemic hyperosmolar state (HSS) or combined DKA and HHS.



Treatment and outcomes

- Insulin was initiated in 98/192 (51%) cases and metformin in 91/192 (47%) cases.
- Diet/lifestyle modification counselling was provided in 126/192 (66%) cases.

Study limitations

- As with any voluntary reporting surveillance system, the CPSP recognizes that all new cases of paediatric NT1DM in Canada may not be captured; therefore, a minimum incidence estimate for NT1DM and its subtypes will be calculated.
- Case-level surveillance data are extracted from patient medical charts following the clinical encounter; therefore, some data elements not collected as part of routine care may be absent from the surveillance totals and, in some cases, insufficient clinical information

is provided to accurately assign a diagnosis of diabetes subtype. However, surveillance serves an important purpose and provides rich clinical data that will allow for a better understanding of Canadian-specific epidemiology and the impact of T2D, MID, and monogenic diabetes in Canadian children.



Conclusions

- With an average of 22 cases of NT1DM reported each month in 2018, it appears that the incidence of this disease, once thought to be rare in children, is on the rise.
- Just over half (98/189, 52%) of NT1DM cases were asymptomatic.
- If reporting rates remain steady, approximately 550 cases of NT1DM will be reported by the end of the surveillance period (May 2019), with the majority of these cases being childhood-onset T2D.



Anticipated study impact

- Study results will provide minimum incidence rates and trends in childhood-onset NT1DM and its subtypes based on Canada's unique ethnic, cultural, and geographic characteristics.
- Results will help define childhood-onset T2D risk factors, clinical characteristics, and diabetes-related complications, as well as differences in approaches to treatment across Canada.
- By largely replicating the first CPSP study on NT1DM, this subsequent study can help determine whether the 'face' of childhood-onset T2D is changing related to demographics, clinical presentation, and severity — information that is critical to designing prevention and treatment programs that meet the specific needs of the populations affected.



Publication and dissemination

Non-type 1 diabetes. Amed S, Sellers E. Canadian Paediatric Society Annual Conference, Quebec City, in May 2018 (oral presentation)

Infantile and later-onset paediatric Pompe disease (glycogen storage disease type II)

October 2017 to September 2019



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? Question

- What is the clinical presentation of infantile and later-onset paediatric Pompe disease in Canada?
- What are the minimum incidence and minimum prevalence of infantile-onset and juvenile-onset paediatric Pompe disease in Canadian children and adolescents?

! Importance

- The classic manifestations in congenital and adult-onset Pompe disease have been well characterized, but it is critical to understand the full spectrum of symptoms and clinical characteristics associated with infantile and juvenile-onset paediatric Pompe disease in order to start prompt management and treatment.
- The incidence and prevalence of infantile and juvenile-onset paediatric Pompe disease in Canadian children and adolescents are unknown.
- Raising awareness among Canadian paediatricians about infantile and juvenile-onset Pompe disease in Canadian children and adolescents is important to ensure that the disease is considered appropriately in the differential diagnosis for children presenting with proximal weakness, hypotonia, respiratory insufficiency, and/or high serum creatine kinase.
- Early diagnosis and treatment are critical for infants with Pompe disease. Study results may provide evidence to support the addition of Pompe disease to the list of diseases for newborn screening.

Methodology

The complete protocol can be accessed at www.cpsp.cps.ca/surveillance.

Case definition

A patient (new or previously diagnosed) of less than 18 years old meeting the following criteria:

1. **Genetic criteria:** Pathogenic mutations affecting both GAA genes (encodes the acid alpha-glucosidase protein) as determined by sequence analysis or deletion/duplication analysis

AND/OR

2. **Biochemical criteria:** Measurement of acid alpha-glucosidase (GAA) enzyme activity performed on one or more of:

- Dried blood spot GAA enzyme activity assay
- Whole blood GAA enzyme activity assay
- Skin biopsy (fibroblast culture) GAA enzyme activity assay
- Muscle biopsy GAA enzyme activity assay

Exclusion criteria

Clinical evidence of proximal muscle weakness without genetic or biochemical confirmation of disease

Unique to this study

Although Canadian Paediatric Surveillance Program (CPSP) studies classically capture minimum incidence rates, this study also aims to capture the prevalence of infantile and later-onset paediatric Pompe cases in Canada.

Results – January to December 2018

TABLE 1 – Infantile and later-onset Pompe cases in 2018

Reported	Reported from Quebec August to December	Duplicates	Excluded	Pending	Met case definition
13	0	0	0	3	10

Cases that met the case definition

- At the time of analysis, 10 paediatric Pompe cases were verified as meeting the case definition in 2018.
- To date, there have been no reported incident cases. Canadian regional laboratories responsible for testing for Pompe disease have been approached for case validation and confirm that no recent positive Pompe enzyme assays have been obtained.

Demographics

- Patient sex was male in the majority of cases in 2018.
- Age ranges of cases were as follows: 7 (70%) were aged 0 to 8 years and the remaining cases were aged 9 to 18 years.
- The geographic distribution of cases was: 6 (60%) from Central Canada and the remainder from Western Canada.
- Population groups were as follows: 6 (60%) cases were Caucasian/White and the remaining cases were from other population groups.
- In 8 (80%) cases the reported child was the first known case in the family.

Presentation

- Decreased enzyme activity was reported for 7 (70%) cases and the diagnosis was done by metabolic centres in Canada.
- The reported mutations for the GAA gene were available for 6 (60%) cases.
- The age range of symptom onset is 0 to 156 months. Six (60%) cases presented with symptoms before 12 months of age.
- Frequent signs or symptoms prior to confirming the diagnosis included the following: 8 (80%) with hypotonia, 5 (50%) with cardiomyopathy, and 5 (50%) with proximal weakness of the arms or proximal weakness of the legs. Other signs and symptoms included: never being able to crawl, stand, or walk; feeding difficulties; enlarged tongue; and failure to thrive.

Treatment and outcomes

- Enzyme replacement therapy was available for all 10 (100%) reported cases after diagnosis. Six (60%) patients who underwent therapy reported clinical improvements.

Study limitations

- As with any voluntary reporting surveillance system, the CPSP recognizes that reporting on minimum incidence rates and/or prevalence rates, can have limitations, including under-representation of the disease/condition in the population. It is possible that some groups of children were missed, for example, those who live in rural or remote areas, as they may be less likely to receive timely specialist care.
- Case-level surveillance data are extracted from patient charts following the clinical encounter. Data elements, for example signs and symptoms prior to diagnosis, laboratory results, and relevant components of the diagnostic assessment may not have been available at the point of care and therefore may be absent from the surveillance totals. However, surveillance serves an important purpose and provides rich clinical data that will allow us to better understand the Canadian-specific epidemiology and the impact of infantile and later-onset Pompe disease.

Conclusions

- In 2018, 10 cases of Pompe disease met the case definition, which is much lower than the anticipated annual number of cases.
- All 10 cases were prevalent cases (diagnosis already known by the physician prior to the launch of the study) and no new incident cases have been reported to date.

Anticipated study impact

Knowledge translation will include comparative data of minimum disease prevalence against what is reported in the literature. Information will be distributed to relevant knowledge users across Canada.

Acknowledgements

We would like to thank Rhiannon Hicks, BSc for her administrative assistance with the study.

Medically serious self-harm in youth requiring ICU admission

January 2017 to December 2018 – Final report



Daphne Korczak

Principal investigator

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Co-investigators

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Collaborator: Yao X

Question

- What is the minimum incidence rate of children and adolescents (less than 18 years of age) admitted to the intensive care unit (ICU) for medically serious self-inflicted injury?
- What are the patterns of presentation, including demographics and psychiatric and medical history, encountered in these cases?

Importance

- Suicide is the second leading cause of death among Canadian youth (15 to 19 years of age), representing more than a quarter of all deaths in this age group in 2015.
- For every adolescent that dies by suicide, it is estimated there are 20 to 40 suicide attempts. However, there is little information available regarding the suicide attempts.
- Youth who make near-fatal suicide attempts – such as those requiring ICU-level care – may closely approximate those who die by suicide, highlighting the need to further understand these suicide attempts.

Methodology

The complete protocol can be accessed at www.cpsp.cps.ca/surveillance.

Case definition

A new patient less than 18 years of age (up to the 18th birthday) meeting **BOTH** of the following criteria:

1. A confirmed or suspected self-harm or suicide attempt (any form of self-poisoning or self-injury regardless of the degree of intent to die)
AND
2. Admitted to an intensive care unit at any time during a hospital admission (for any duration)

Exclusion criteria

Accidental poisoning (e.g., intoxication) or injury

Results – January 2017 to December 2018

TABLE 1 – Medically serious self-harm cases from January 1, 2017 to December 31, 2018

Year	Reported	Reported from Quebec August to December 2018	Duplicates	Excluded	Pending	Met case definition
2017	80	—	0	5	17	58
2018	60	4	3	0	17	36
Total	140	4	3	5	34	94

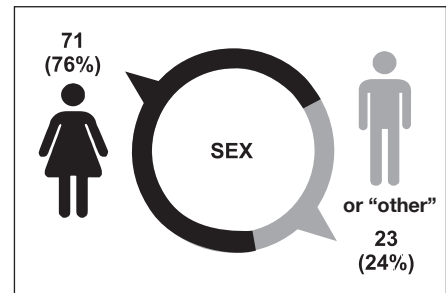
Cases that met the case definition

- At the time of analysis in January 2019, 94 cases met the case definition over the study period, of which 87 (93%) were confirmed self-harm and 7 (7%) were suspected self-harm.

Demographics

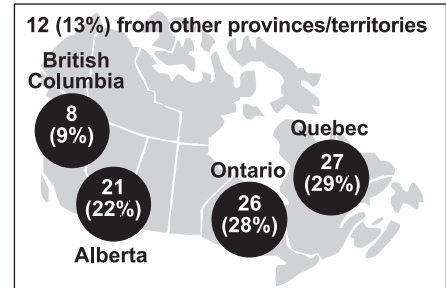
- Patient sex was female in three quarters of these adolescents. There were 71 (76%) females and 23 (24%) males or “other.”
- The mean age of the cases was 15.2 years (range: 11.2–17.9).

- The geographic distribution of cases was: 27 (29%) from Quebec, 26 (28%) from Ontario, 21 (22%) from Alberta, 8 (9%) from British Columbia, and 12 (13%) from other provinces/territories
- Population groups were indicated for 84 cases with the majority being from the following groups: 61 (73%) White, 11 (13%) Asian, and 8 (10%) Aboriginal with the remaining cases being from other or multiple groups.
- The majority (69/74, 93%) of the youth were born in Canada.
- Seventy-nine percent (72/91) of the youth were living with their biological parent(s): 51% (46/91) with one parent and 29% (26/91) with both parents.



Presentation

- Among females, the most common method of self-inflicted injury was overdose by medication ingestion (54/71, 76%) compared to hanging among males (9/22, 41%).
- The most common precipitating events associated with suicide attempts (both fatal and non-fatal) were the following (one case may be associated with multiple events): family conflict was with 43/94 (46%) cases, romantic relationship crisis with 20/94 (21%) cases, and peer conflict with 19/94 (20%) cases.
- Of the 44 cases with a previous suicide attempt(s), 84% (37/44) were female.
- A higher proportion of females (50/65, 77%) than males (11/20, 55%) had received a psychiatric diagnosis in the past.
- A higher proportion of females (44/64, 69%) than males (6/20, 30%) were under the care of a psychiatrist or other mental health professional at the time of ICU admission.
- In 32% (22/68) of cases, the parents/caregivers were aware that their child was considering suicide.



Treatment and outcomes

- During the ICU admission, 47% (44/93) of youth received ventilation, 29% (25/87) received an antidote, and 27% (24/88) received hemodynamic support as part of treatment. Five or fewer cases required each of dialysis, surgery, or exchange transfusion.
- Of the 82 cases that were known to be discharged from the ICU, 11% (8/75) had permanent impairment from the episode of self-harm.
- Of the 82 cases that were known to be discharged from the ICU, 93% (76) were known to be referred for follow-up with a psychiatrist and/or a mental health professional.
- There were 11 deaths by suicide (11/94, 12%) reported. In this sample, more males than females died by suicide. Nine (82%) deaths were the result of hanging.

Study limitations

- As with any voluntary reporting surveillance system, the Canadian Paediatric Surveillance Program recognizes that reporting on minimum incidence rates can have limitations, including under-representation of the disease in the population. It is possible that some groups of children were missed, for example those who live in rural or remote areas or adolescents treated in adult settings.
- Case-level surveillance data are extracted from patient charts following the clinical encounter. Data elements, such as psychiatric or family history, may not have been available at the point of care and therefore may be absent from surveillance totals. However, surveillance serves an important purpose and provides rich clinical data that will allow for a better understanding of medically serious self-harm in youth requiring ICU admission.



Conclusions

- These findings are consistent with epidemiologic data that observe a gender paradox in youth suicide in which females demonstrate a higher rate of mental health care use and suicide attempts, while males display decreased mental health care engagement and a higher rate of suicide mortality. Future research in suicide prevention should consider both gender and study setting to meaningfully inform further risk factor identification and effective intervention design.
- Findings are also consistent with previous research reporting that family conflict is a salient risk factor for suicide among youth, and this is a potential target for suicide prevention intervention.



Anticipated study impact

The present study seeks to extend current knowledge by providing greater detail on the personal and psychiatric history, situational factors, and management of adolescents following a near-fatal suicide attempt.



Publication and dissemination

Serious self-harm requiring intensive care unit admission: Understanding near-fatal suicide attempts. Korczak DJ, Skinner R, Dopko R. *Paediatr Child Health* 2018 Jul 25; pxy077, <https://doi.org/10.1093/pch/pxy077> [Epub ahead of print]

Suicide risk among children and adolescents: Making evidence-informed decisions. Korczak D. Canadian Paediatric Society Annual Conference, Quebec City, in May 2018 (oral presentation)

Ophthalmia neonatorum caused by *N gonorrhoeae* or *C trachomatis*

November 2018 to October 2020



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Principal investigators

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? Question

- What is the minimum incidence of ophthalmia neonatorum (ON) caused by *Neisseria gonorrhoeae* or *Chlamydia trachomatis* in Canada?
- Do rates of ON caused by *N gonorrhoeae* or *C trachomatis* differ in jurisdictions with mandatory ocular prophylaxis versus those without?

! Importance

- ON is neonatal conjunctivitis that occurs within the first month of life. *C trachomatis* and *N gonorrhoeae* have been reported to account for up to 40% and 1% of ON cases respectively.
- Without preventive measures, gonococcal ophthalmia neonatorum (GON) occurs in 30% of infants exposed during delivery; without treatment, the disease may progress rapidly and cause severe consequences. Infants born to women with untreated chlamydia infection at delivery have a 30% to 50% risk of developing chlamydial ophthalmia neonatorum (CON).
- Ocular prophylaxis for ON with erythromycin is mandated in some provinces.
- In 2015, a Canadian Paediatric Society position statement recommended the discontinuation of mandatory ocular prophylaxis for ON because of the questionable efficacy of erythromycin. The position statement advocated for the enhancement of routine sexually transmitted infection (STI) screening and treatment programs for pregnant women.
- Concerns about the current effectiveness of STI screening and treatment programs for pregnant women include the worry that discontinuation of erythromycin ocular prophylaxis could result in increased rates of GON and CON.
- ON is no longer a nationally notifiable disease which has raised concerns about the ability to monitor the effect of changing policies on rates of ON. Gonorrhea and chlamydia infections in children less than 1 year of age are still notifiable through the National Disease Surveillance System (NDSS).

➡ Methodology

The complete protocol can be accessed at www.cpsp.cps.ca/surveillance.

Case definition

Any patient less than 28 days of age (4 weeks) at onset of symptoms, with clinical features of ophthalmia neonatorum including at least one of the following:

- Conjunctival/ocular erythema
- Conjunctival/ocular discharge
- Conjunctival and/or peri-ocular swelling

AND

N gonorrhoeae isolated in culture or identified by nucleic acid amplification test in specimens from the eye, blood, CSF, or other sterile site

OR

C trachomatis isolated in culture or identified by nucleic acid amplification test in specimens from the eye, nasopharynx, or other respiratory tract specimen

Exclusion criteria

- Positive microbiology test for *C trachomatis* or *N gonorrhoeae* without any associated clinical abnormality
- Ophthalmia neonatorum associated with another microorganism



Results – November to December 2018

TABLE 1 – ON cases from November 1 to December 31, 2018

Reported	Reported from Quebec August to December	Duplicates	Excluded	Pending	Met case definition
<5	0	<5	<5	<5	<5

Cases that met the case definition

- Fewer than five cases of ON were verified as meeting the case definition in the first two months of data collection.

Demographics

- As per CPSP policy, case numbers and data for fewer than five cases cannot be presented.

Presentation, treatment, and outcomes

While specific information on this study cannot be presented at the current time due to small case numbers, available literature demonstrates that:

- If left untreated, ON caused by *N gonorrhoeae* can lead to corneal ulceration, ocular perforation, or panophthalmitis (rarely).
- The clinical manifestations of ON caused by *C trachomatis* vary from mild conjunctival infection with scant watery discharge to severe mucopurulent discharge with eyelid edema, chemosis, and pseudomembrane formation. If left untreated, superficial corneal vascularization and conjunctival scarring can occur.
- Currently, erythromycin is the only product marketed in Canada for neonatal ocular prophylaxis.

Study limitations

- As with any voluntary reporting surveillance system, the CPSP recognizes that reporting on minimum incidence rates can have limitations, including under-representation of the disease/condition in the population. It is possible that some groups of children were missed, for example, those who live in rural or remote areas, as they may be less likely to receive timely specialist care.
- Case-level surveillance data are extracted from patient medical charts following the clinical encounter. Certain data elements may not have been available at the point of care and therefore may be absent from surveillance totals. Surveillance still serves a very important purpose by providing rich clinical data that allow for a better understanding of the diagnosis, treatment, and preventive approaches of ON caused by *N gonorrhoeae* or *C trachomatis*.



Conclusions

- This CPSP study will continue until October 2020 and monitor the number of GON and CON cases that are reported to the program. Case numbers will be compared to case numbers collected via the NDSS.
- Data analysis will include the proportion of infections occurring in infants of mothers who were screened during pregnancy, in infants of mothers who were treated in pregnancy, and in infants who received ocular prophylaxis.



Anticipated study impact

- This study will provide valuable clinical and epidemiological information on cases of ON across Canada, as ON is no longer a nationally notifiable disease.
- Information on infection rates can be used to understand the effect of current neonatal ocular prophylaxis practices on disease rates, and may inform future public health policy changes.

Rh sensitization

June 2016 to May 2018 – Final report



Michael Sgro

Principal investigator

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Co-investigators

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? Question

What is the burden of Rh sensitization and Rh disease-associated neonatal severe hyperbilirubinemia in Canada?

! Importance

- Rh sensitization occurs when women whose red blood cells are Rh(D)-antigen negative develop anti-Rh(D) antibodies either during a previous pregnancy in which the fetus is Rh(D) positive or by exposure to Rh antigens from blood products/transfusion.
- Neonates born to Rh-sensitized mothers may present with severe jaundice, anemia, and death from acute or chronic bilirubin encephalopathy or brain damage resulting from severe neonatal hyperbilirubinemia.
- Rh disease is now considered rare in countries where Rh prophylaxis is used and the blood type and Rh sensitization status of the mother is usually known at the time of a delivery.

➔ Methodology

The complete protocol can be accessed at www.cpsp.cps.ca/surveillance.

Case definition

Any infant 60 days of age or less with Rh(D) sensitization fulfilling ALL of the following criteria:

- Mother is Rh negative (D-negative)
- Mother has positive antibody screen due to anti-D. (This must be a maternal allo-anti-D, not passive anti-D from Rh(D) immunoglobulin (RhoGAM))
- Cord or infant blood group is Rh positive (D-positive)

✓ Results – June 2016 to May 2018

TABLE 1 – Rh sensitization cases from June 1, 2016 to May 31, 2018

Year	Reported	Reported from Quebec August to December 2018	Duplicates	Excluded	Pending	Met case definition
2016*	26	—	0	1	8	17
2017	46	—	1	1	12	32
2018†	17	—	2	0	2	13
Total	89	—	3	2	22	62

* June 1 to December 31, 2016

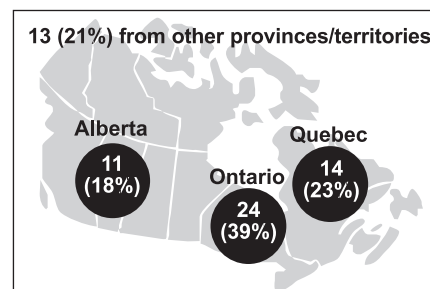
† January 1 to May 31, 2018

Cases that met the case definition

- At the time of analysis, 62 cases of Rh sensitization were verified as meeting the case definition over the study period.

Demographics

- The geographic distribution of cases was: 11 (18%) from Alberta, 24 (39%) from Ontario, 14 (23%) from Quebec, and 13 (21%) from other provinces/territories.
- Maternal country of birth was reported as Canada in 42 (68%) cases. The remaining cases were born to mothers from India, Russia, Kenya, Nigeria, Nicaragua, Afghanistan, Armenia, Syria, Bangladesh, and the Philippines, where Rh immune globulin (RhIg) may not be routinely available, or the maternal country of birth was unknown.



Presentation

- The median gestational age was 37 weeks (range: 35–40 weeks).
- The median age at presentation was 2 hours.
- The median hemoglobin at presentation was 137.5 g/L (range: 33–203 g/L).
- The median peak micro bilirubin (MBR) level was 280 µmol/L (range: 92–771 µmol/L).

Treatment and outcomes

- The median duration of phototherapy (in hours) was 139.0.
- Nine (15%) cases received one or more exchange transfusions.
- Thirty (48%) cases received IVIG (1–5 doses, median 3 doses).
- Thirteen (21%) cases received packed red blood cell transfusion (1–3 transfusions, median 1 transfusion).
- Six (10%) infants presented with acute bilirubin encephalopathy, and fewer than five presented with seizures.

Study limitations

- The study was not able to capture cases of Rh(D) sensitization in a mother if the infant was asymptomatic.
- As with any voluntary reporting surveillance system, the CPSP recognizes that reporting on minimum incidence rates can have limitations, including under-representation of the disease/condition in the population.
- Case-level surveillance data was extracted from patient charts following the clinical encounter. Data elements, such as pending test results or family history, may not have been available at the point of care and therefore may be absent from the surveillance totals. However, surveillance serves an important purpose and provides rich clinical data that will allow us to better understand Rh sensitization.

Conclusions

- Rh disease continues to exist in Canada.
- Despite current public health measures in place across the country, additional work must be done to raise awareness of Rh disease, to prevent the disease whenever possible, and to minimize sequelae when it does occur.
- The immigration of women to Canada who had prior pregnancies in countries where RhIg is not routinely available was likely a factor in some cases of Rh(D) sensitization.
- The ongoing burden of Rh disease worldwide, as well as the possibility of emerging RhIg refusal, should be considered in future awareness and prevention efforts.

Anticipated study impact

- The study identified potential risk factors associated with Rh(D) sensitization among pregnant women in Canada, including immigration of women to Canada who had prior pregnancies in countries where RhIg is not routinely available. The global burden of Rh(D) sensitization has been well described, and global efforts towards eradication are ongoing.
- There is a need for optimization of our national RhIg program to ensure avoidable causes of sensitization are mitigated.
- The study highlighted the burden of Rh disease in Canada, demonstrating the need for increased awareness of Rh disease and education for patients, health care providers, and policy makers to optimize prevention and the care of affected infants.

Publication and dissemination

Rh sensitization in Canada is not obsolete. Baker JM, Campbell DM, Bhutani VK, Sgro M. *Paediatr Child Health* 2017;22(4):238–9

Rh sensitization: A two-year Canadian national surveillance study. Baker JM, Campbell D, Pavenski K, Decou ML, Hollamby K, Jegathesan T, Bhutani V, Zipursky A, Sgro M. The Hospital for Sick Children Haematology Research Day, Toronto, in November 2018 (oral presentation)

Canadian Infants Affected by Rh Sensitization: A 2-year National Surveillance Study. Baker JM. University of Toronto, Divisions of Hematology & Hematopathology 12th Annual Academic Day, Toronto, in April 2019 (poster presentation)

Canadian Infants Affected by Rh Sensitization: A 2-year National Surveillance Study. Gnanalingam A. University of Toronto, 18th Annual Neonatal Research Day, Toronto, in April 2019 (poster presentation)

Infants affected by Rh sensitization: A two-year Canadian national surveillance study. Baker JM, Campbell D, Pavenski K, Decou ML, Hollamby K, Jegathesan T, Bhutani V, Zipursky A, Sgro M. Canadian Paediatric Society Annual Conference, Toronto, in June 2019 (poster presentation)

Acknowledgements

The investigators would like to thank Aidan Campbell for the assistance with the data entry and Aasha Gnanalingam, BSc (MPH, Class of 2020) for compiling the report.

Serious and life-threatening events associated with non-medical (recreational) cannabis use in Canadian children and youth

September 2018 to October 2020



Richard E. Bélanger

Principal investigators

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Collaborator: Dirk Huyer, MD, Chief Coroner for Ontario

? Question

- What is the minimum incidence of serious and life-threatening events associated with non-medical cannabis use in Canadian children and youth?
- What are the clinical presentations and associated medical needs of children and youth presenting with serious and life-threatening events related to non-medical cannabis exposure?
- Are there changes in the incidence of serious and life-threatening events during the two-year time period following cannabis legalization?

! Importance

- There is currently no scientific data describing the impact of cannabis legalization on the health of Canadian children and youth.
- Data provided by this study will be used to assess the impact of cannabis legalization in the paediatric population.

↻ Methodology

The complete protocol can be accessed at www.cpsp.cps.ca/surveillance.

Case definition

Any child or adolescent less than 18 years of age (up to the 18th birthday) presenting with a new health condition or a deteriorating chronic/previously diagnosed condition resulting in either hospitalization (inpatient, intensive care unit, psychiatric), permanent disability, or death, which was likely primarily caused by the use of cannabis for non-medical (recreational) purposes.

This includes either intentional or unintentional exposure to cannabis in a child or adolescent, or a condition resulting from use by another individual, such as a friend or a parent/caregiver, who is under the influence of cannabis.

Exclusion criteria

- A condition resulting from cannabis use for non-medical purposes during pregnancy/breastfeeding
- A condition resulting from cannabis use for medical purposes

✓ Results – September to December 2018

TABLE 1 – Serious and life-threatening events associated with non-medical cannabis use cases from September 1 to December 31, 2018

Reported	Reported from Quebec August to December	Duplicates	Excluded	Pending	Met case definition
16	3	0	0	2	11

Cases that met the case definition

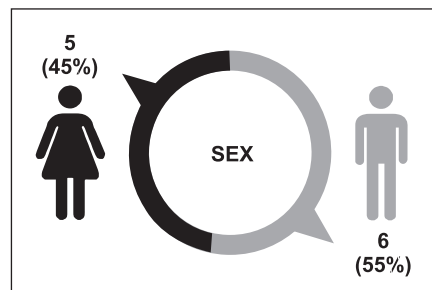
- In total, 16 cases of cannabis related exposure were reported through the CPSP network between September and December 2018.
- At the time of analysis, 11 cases of serious and life-threatening events associated with non-medical cannabis use were verified as meeting the case definition in 2018.

Demographics

- Patient sex was female in 5 (45%, 95% CI 16–74) cases and male in 6 (55%, 95% CI 25–83) cases.
- Cases ranged in age from less than 1 year to 17 years, with a mean of 9 years and a median of 7 years.

Presentation

- Eighty-two percent (9/11, 95% CI 59–100) of cases were confirmed to be cannabis-related exposures through positive urine qualitative testing.
- In 64% (7/11, 95% CI 35–92) of cases exposure was unintentional.
- The majority (6/7, 86%; 95% CI 25–84) of unintentional exposures involved ingestion of cannabis edibles (gummies, chocolate, candy). All of these cases consumed edible cannabis belonging to a parent/caregiver or grandparent.



Treatment and outcomes

- All 11 (100%) cases were hospitalized; 73% (8/11, 95% CI 46–99) of these received physical treatment (e.g., observation in hospital, anti-emetics, catheter).
- Forty-five percent (5/11, 95% CI 16–75) of cases received a referral to a mental health or psychosocial assistance professional (e.g., social worker).

Study limitations

- Reporting on minimum incidence rates can have limitations, including under-representation of the disease in the population. It is possible that some groups of children will be missed, for example, those who live in rural or remote areas, as they may be less likely to receive timely specialist care. Youth who are approaching transition-to-adult-care age may also be under-represented, as they may be treated by an adult provider in an adult facility.
- Moreover, case-level surveillance data is extracted from patient charts following the clinical encounter. Data elements, including details of history, physical examination, and relevant components of the diagnostic assessment not collected as part of routine care will be absent from the surveillance totals.

Conclusions

In the first four months of the study, it was notable that 64% (7/11, 95% CI 35–92) of cannabis exposures were unintentional, and that the vast majority of unintentional exposures were related to the ingestion of edibles such as gummies, chocolate, and candy. All of these cases consumed edible cannabis belonging to a parent/caregiver or grandparent. This early trend will be monitored as the study continues.

Anticipated study impact

- This study will provide Canadian-specific data on the impact of cannabis legalization on the health and well-being of children and youth. This data may be used to inform policies and further regulation related to non-medical cannabis.
- The information from this study may be adapted for public education materials.

Severe microcephaly

June 2016 to May 2018 – Final report



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Co-investigators

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Collaborators: Evans J, Tataryn J

? Question

What is the minimum incidence of severe microcephaly in Canada and what is the spectrum of etiology and clinical manifestations seen in these infants?

! Importance

- Congenital microcephaly is an anomaly of the central nervous system that begins *in utero*. It is a condition in which an infant's head is significantly smaller than the heads of other children of the same age and sex at the time of birth.
- In 2015, there was an increase in the number of microcephaly cases reported in Brazil. The increased incidence in Brazil, followed by similar experiences in other countries, was subsequently linked to an outbreak of Zika virus, where pregnant women infected with Zika were found to give birth to infants with severe microcephaly and other congenital abnormalities.
- Given the frequent travel of Canadians to Zika-endemic regions, it was critical to monitor for microcephaly in Canada, both to establish baseline rates and to understand the potential association with Zika virus infection.
- Surveillance for severe microcephaly in Canada through the Canadian Paediatric Surveillance Program (CPSP) started in June of 2016. This project is complementary to the CPSP surveillance for congenital Zika syndrome (CZS) launched in March 2017.

➡ Methodology

The complete protocol can be accessed at www.cpsp.cps.ca/surveillance.

Case definition

Any new patient less than 12 months of age, with a head circumference measurement less than three standard deviations below the mean (0.13th centile) for gestational age and sex, based on the sex-specific World Health Organization growth parameters:

- Female term infant with a head circumference of less than 30.3 cm
- Male term infant with a head circumference of less than 30.7 cm
- Preterm infant (less than 38 weeks' gestation), as per INTERGROWTH-21st study standards

Unique to this study

Given the significant international interest in this issue, the CPSP has partnered with the International Network of Paediatric Surveillance Units to align research questions and data definitions. Parallel studies are ongoing in the United Kingdom, Australia, and New Zealand, and multi-national data will be collated and compared at the completion of this surveillance project. This partnership represents the largest international research collaborative investigating the epidemiology of severe microcephaly.

✓ Results – June 2016 to May 2018

TABLE 1 – Severe microcephaly cases from June 1, 2016 to May 31, 2018

Year	Reported	Reported from Quebec August to December 2018	Duplicates	Excluded	Pending	Met case definition
2016*	30	—	0	12	2	16
2017	40	—	1	14	5	20
2018†	10	—	0	3	4	3
Total	80	—	1	29	11	39

* June 1 to December 31, 2016

† January 1 to May 31, 2018

Cases that met the case definition

- At the time of analysis, 39 cases of severe microcephaly were verified as meeting the case definition over the 24 months of the study.
- All cases included in this study were live births and did not include stillbirths, therapeutic abortions, or intra-uterine fetal deaths.

Demographics

- Patient sex was female in 17 (44%) cases, male in 21 (54%) cases, and 1 was unspecified.
- The mean gestational age at birth was 38 weeks (2.4 SD) and the mean weight at birth was 2151 grams (932 SD).
- The geographic distribution of cases was: 16 (41%) from Ontario, 9 (23%) from Quebec, 5 (13%) from Nova Scotia, and 9 (23%) from other provinces/territories.

Presentation, treatment and outcomes

- The majority (36, 92%) of infants were singleton births.
- The average head circumference at birth was 28.9 cm (2.0 SD).
- Seven (18%) cases had a confirmed genetic disorder identified. For the remaining 32 cases, the leading suspected causes of severe microcephaly varied and included: 7 (22%) genetic causes and five or fewer cases each of ischemia, infections, and unknown or other causes.
- Table 2 displays other common features identified in severe microcephaly babies and emphasizes that those with severe microcephaly may present with other significant clinical features.
- Nine (23%) mothers reported travelling outside of Canada during pregnancy or in the three months prior to conception, with 6 (67%) of these mothers having travelled to Zika-infected areas.
- Zika infection was suspected as the cause of microcephaly in all nine cases associated with a report of maternal travel during pregnancy.
- Zika was investigated as a potential cause of severe microcephaly for five of the six mothers who travelled to Zika-endemic countries and the remaining case did not specify.
- Fewer than five cases reported a positive Zika virus laboratory confirmation; however, results of some of the laboratory tests were not reported.

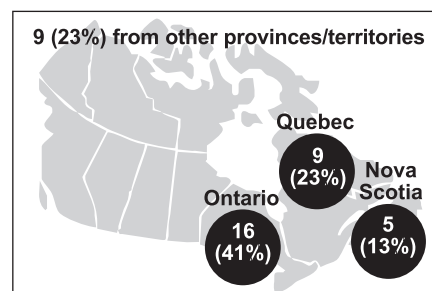
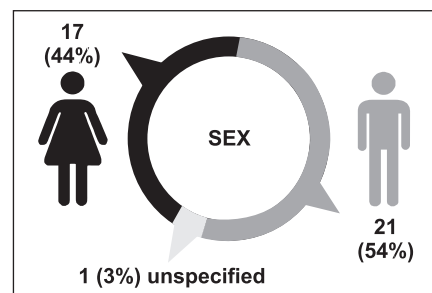


TABLE 2 – Most common clinical features seen in severe microcephalic infants (n > 5)

Clinical features	Percent (%)
Dysmorphism	41
Intracranial abnormalities	41
Craniofacial disproportion	23
Spasticity	18
Hypotonia	18
Eye abnormalities	15

Study limitations

- The case definition of severe microcephaly is restrictive and therefore only the most serious cases of the condition were captured.
- This study captured only live-born infants. This underestimates the frequency of severe microcephaly cases, as stillbirths, intra-uterine fetal deaths, and abortions affected by severe microcephaly would not be captured.
- As with any voluntary reporting surveillance system, the CPSP recognizes that reporting on minimum incidence rates can have limitations, including under-representation of the disease/condition in the population. It is possible that some groups of babies were missed, for example those who live in rural or remote areas, as they may be less likely to receive timely specialist care.
- Case-level surveillance data are extracted from patient charts following the clinical encounter. Data elements, such as pending laboratory results or components of the diagnostic assessment, may not have been available at the point of care and therefore may be absent from the surveillance totals.

Conclusions

- The minimum estimated incidence of severe microcephaly in Canada for the two-year period of surveillance was 5.1 per 100,000 births or 1 in 19,689 births (based on Statistics Canada Estimates of Births).
- The causes of severe microcephaly are varied and are often not determined. The majority of cases with known cause are genetic.
- Zika was investigated as a potential cause for most cases associated with maternal travel to a Zika endemic country.
- Fewer than five cases of Zika-associated severe microcephaly have been confirmed in Canada.

Anticipated study impact

There are no case-level national data on microcephaly in Canada. This study will provide valuable clinical and epidemiological information on severe microcephaly.

Publication and dissemination

Small bite, big problem: Understanding severe microcephaly in Canada. Nelson CRM, Demarsh A, Miller SP, Morris SK, Moore Hepburn C, Bitnun A, Moore A, Shevell M, Evans J, Tataryn J. *Paediatr Child Health* 2017; 22(8):504–5

Severe microcephaly in Canada. Nelson C. Canadian Paediatric Society Annual Conference, Quebec City, in May 2018 (poster presentation)

Acknowledgements

The study team acknowledges and thanks Mihaela Gheorghe and Matthew Krupovich (Public Health Agency of Canada) for their contributions to data entry and analysis.

Severe obesity and global developmental delay in preschool children

February 2018 to January 2020



Catherine Birken

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? Question

- What is the minimum incidence of severe obesity (SO) and global developmental delay (GDD) in preschool children in Canada?
- What are the age of onset, risk factors, and health care utilization associated with SO and GDD in Canadian preschoolers?

! Importance

- To date, no Canadian studies have examined co-morbid SO and GDD in children.
- Understanding the incidence and risk factors of SO and GDD is necessary to best direct appropriate investigations and to develop effective management strategies.

↻ Methodology

The complete protocol can be accessed at www.cpsp.cps.ca/surveillance.

Case definition

Any new case of a child ≤ 5 years of age with:

1. Severe obesity, defined as body mass index ≥ 99.9 th percentile according to references developed by the World Health Organization and the Canadian Pediatric Endocrine Group. The absolute cut-offs by age and sex can be accessed in the study protocol at www.cpsp.cps.ca/surveillance.

AND

2. Global developmental delay, defined as a significant delay in two or more developmental domains, including:
 - Gross motor
 - Fine motor
 - Speech/language
 - Cognitive
 - Social/personal
 - Delay in activities of daily living

Unique to this study

An infographic was created to raise awareness of the study and was disseminated through existing research and clinical networks (e.g., Team to Address Bariatric Care in Canadian Children, Women and Children's Health Research Institute, Maternal Infant Child and Youth Research Network). The infographic was designed to encourage CPSP participants to report cases meeting the case definition.

✓ Results – February to December 2018

TABLE 1 – SO and GDD cases from February 1 to December 31, 2018

Reported	Reported from Quebec August to December	Duplicates	Excluded	Pending	Met case definition
37	3	1	3	19	11

Cases that met the case definition

- At the time of analysis, 11 cases of co-morbid SO and GDD were verified as meeting the case definition in 2018.

Demographics

- The mean age of the cases was 3.49 years (SD 1.18).
- The geographic distribution of cases was: 7 (64%) from Ontario and the remaining cases from other provinces.
- Population groups were as follows: 5 (46%) cases were White and the remaining cases were First Nations, South Asian, Latin American, and Southwest Asian.

Presentation

- Cases had a mean body mass index z-score of 4.2 (SD 1.3).
- The mean age of GDD diagnosis was 2.3 years (SD 1.0).
- The mean age of first weight concern was 2.7 years (SD 1.3).
- Health problems reported were snoring, asthma, nutrient deficiencies, recurrent otitis media, attention deficit disorder, and fatty liver disease.

Treatment and outcomes

- Genetic tests were ordered for 8 (73%) cases, including 7 (64%) microarray.
- Central nervous system imaging was ordered in fewer than five cases.
- Dietitians were involved in the care of 10 (91%) cases and developmental paediatricians were involved in the care of 7 (64%) cases.
- The most significant challenges reported in caring for children with SO and GDD included a lack of developmental and obesity health services and the wait time between referral and program access.

Study limitations

- As with any voluntary reporting surveillance system, the CPSP recognizes that reporting on minimum incidence rates can have limitations, including under-representation of the disease in the population. It is possible that some groups of children were missed, for example those who live in rural or remote areas, as they may be less likely to receive timely specialist care.
- Case-level surveillance data are extracted from patient charts following the clinical encounter. Data elements, such as pending test results or family history, may not have been available at the point of care and therefore may be absent from the surveillance totals.

Conclusions

- To date, concerns about SO among children with GDD are recognized at approximately 2.7 years of age.
- Multidisciplinary service provision was common. However, challenges in access to expert and timely care were identified.

Anticipated study impact

- This study will be the first to establish the minimum incidence of SO and GDD in preschool children in Canada.
- The study will help identify patient demographics, age of onset, risk factors, and health care service use associated with SO and GDD.
- Results will be used to promote early recognition and treatment of these co-morbid conditions, and to improve paediatric providers' awareness of SO and GDD among Canadian preschoolers.

Acknowledgements

We wish to thank Nicole Gehring (University of Alberta) and Stephan Oreskovich (Hospital for Sick Children) for their help with the CPSP study on severe obesity and global developmental delay in preschool children.

One-Time Surveys

Neonatal abstinence syndrome

July 2018



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? Question

- What are the variations in practice in the management of neonatal abstinence syndrome (NAS) across Canada?
- What are Canadian paediatricians' knowledge and perceptions around NAS care?

! Importance

- NAS is a complex, multi-system disorder caused by opioid withdrawal after *in utero* exposure.
- According to the Canadian Institute for Health Information (CIHI), there were approximately 1,850 cases of NAS across the country in 2016–2017. The number represents a significant increase over the last 10 years, and is expected to continue to rise.
- A recent Canadian Paediatric Society (CPS) practice point, *Managing infants born to mothers who have used opioids during pregnancy*, was published in early 2018 to address gaps in knowledge by outlining evidence-based care principles.

↪ Methodology

A one-time survey was sent to paediatricians and paediatric subspecialists through the Canadian Paediatric Surveillance Program (CPSP). The survey tool can be accessed at www.cpsp.cps.ca/surveillance.

✓ Results

The survey response rate was 31% (878/2,808).

Number of NAS cases seen in the prior 12 months

- Respondents reported having monitored the following number of NAS cases in their practice settings in the past 12 months: 28% (87/307) saw 1 to 5 cases, 17% (51/307) saw 6 to 10 cases, 9% (27/307) saw 11 to 15 cases, 7% (21/307) saw 16 to 20 cases, and 22% (68/307) saw 21 or more cases. The remaining respondents reported having monitored no cases of NAS or did not recall the number of cases monitored.

CPS practice point and NAS management guidelines

- Sixty-seven percent (203/302) of respondents were aware of the new CPS practice point.
- Sixty-three percent (170/269) of respondents agreed with the recommendations made in the CPS practice point.
- Sixty-four percent (194/302) of respondents reported that their practice settings have established NAS guidelines and 29% (41/142) were using the CPS practice point as their primary established guideline.

Reported first location of care for stable infants with NAS

- The reported first location of care for stable infants with NAS were as follows: 46% (134/292) were rooming-in with the mother, 30% (88/292) were in the neonatal intensive care unit (NICU), 22% (63/292) were in the special care nursery, and 2% (6/292) were on a paediatric ward.

Attitudes towards rooming-in

- Eighty percent (237/298) of respondents supported rooming-in.
- The leading reasons for not adopting rooming-in included the following: 70% (102/146) said it was due to staff concern regarding monitoring and evaluation, 56% (81/146) stated insufficient nursing resources, and 49% (72/146) stated insufficient education and training of staff.

Variation in NAS care

- Finnegan scoring to assess for NAS severity was used in 51% (149/290) of respondents' centres.
- The most common pharmacological treatment is morphine only, reported by 40% (111/278) of respondents.
- A minimum length of observation of 72 hours following opioid exposure (as recommended by the CPS practice point) is used by 38% (105/276) of respondents; 36% (100/276) observe for longer.
- Sixty-five percent (181/279) of respondents' practice centres do not discharge infants on pharmacological treatment.

Survey limitations

- As the response rate was 31%, these results may not accurately describe the actual national practice variation.
- Paediatricians who commonly care for patients with NAS may have been more likely to respond to the survey, introducing bias into the results.
- Not all respondents provided answers to all of the questions. Thus, denominators vary throughout this summary.



Conclusions

- Despite the publication of a CPS practice point in early 2018, significant variation exists in the management practices for NAS, including first location of care, pharmacological treatment, length of observation, and discharge requirements.
- Less than half of respondents' practice settings employ rooming-in as the first location of care. The largest barriers to the adoption of rooming-in were staff concern over monitoring and evaluation, insufficient nursing resources, and the need for education and training.



Anticipated survey impact

- These results highlight the need for additional education of health care professionals regarding NAS and its management.
- Identified barriers to practice point adoption can inform targeted educational outreach as well as resource allocation to ensure optimal rates of rooming-in nationally.



Publication and dissemination

Neonatal abstinence syndrome: Variation and barriers in care in Canada. Puvitharan D, Dow K, Lacaze-Masmonteil T, Little J, Khurshid F. Pediatric Academic Societies 2019 Meeting, Baltimore, in April 2019 (poster presentation)

Neonatal abstinence syndrome: Variation and barriers in care in Canada. Puvitharan D, Dow K, Lacaze-Masmonteil T, Do M, Nelson C, Little J, Khurshid F. Canadian Paediatric Society Annual Conference, Toronto, in June 2019 (poster presentation)

Acknowledgements

We would like to thank Ms. Melanie Laffin Thibodeau and Dr. Charlotte Moore Hepburn for their contributions, as well as the Public Health Agency of Canada for providing funding to support this work.

Procedural skill needs for Canadian paediatricians: A national profile

March 2018



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? Question

What are the essential procedures a Canadian paediatrician practising general paediatrics should be competent to perform?

! Importance

- The procedural skill set required of Canadian paediatricians in general practice is evolving and the list of procedures contained in the 2008 *Objectives of Training in Pediatrics* issued by the Royal College of Physicians and Surgeons of Canada may require updating.
- Competency-based training, both at the post-graduate and continuing professional levels, should focus on those skills that are necessary for current paediatric practice.
- Recognizing that procedures training is resource-intensive, it is crucial to invest in those skills that are essential for practising professionals. In addition to adequate training, the maintenance of procedural skill competency of paediatricians in general practice is necessary to provide high-quality care for Canadian children.

↪ Methodology

A one-time survey was sent to paediatricians and paediatric subspecialists through the Canadian Paediatric Surveillance Program (CPSP). The survey tool can be accessed at www.cpsp.cps.ca/surveillance.

✓ Results

The survey response rate was 33% (934/2,822). Of the total number of survey respondents, 63% (589/934) reported that they spend some or all of their clinical work time in general paediatrics. The following survey results pertain only to those 589 respondents.

Respondents

- Of the respondents who indicated that they practice general paediatrics, 69% (407/589) practise full-time general paediatrics and 31% (182/589) practise part-time general paediatrics.
- The 589 full- and part-time general paediatricians reported the following practice settings: 52% (309) community office, 41% (243) tertiary care university teaching hospital, 38% (222) community hospital, 11% (65) rural/remote hospital, and 6% (38) indicated another practice setting. Percentages total more than 100% as many participants reported practice in more than one setting.
- Career duration ranged from 23% (133/589) who had been in independent paediatric practice for less than or equal to five years to 42% (245/589) who had been in practice for 20 years or more.
- Seventy-one percent (290/407) of the full-time and 70% (127/182) of the part-time general paediatricians reported that they perform or supervise procedures. The remaining 29% (172/589) of general paediatricians indicated that they never perform or supervise procedures.
- Completed surveys were received from all provinces and territories and were geographically representative of regional population distributions.

Procedure frequency

- Of all the general paediatric respondents, the procedures most frequently performed included the following: 39% (229/589) perform infant bag-valve mask ventilation monthly, 33% (197/589) perform lumbar puncture monthly, and 28% (163/589) perform ear curettage monthly.

- The procedures least frequently performed by respondents included the following: 93% (547/589) never perform PICC placement, 92% (542/589) never perform central venous line (jugular/femoral) insertion, and 88% (516/589) never perform peripheral arterial line insertion.

Essential procedures

- The following procedures were rated as highly essential: 91% (335/367) identified lumbar puncture and 88% (324/369) identified chest compressions.
- The following procedures were rated as least essential: 93% (339/365) identified PICC insertion and 90% (330/367) identified central venous line (jugular/femoral) insertion.

Survey limitations

- Self-reports of the frequency of performing procedural skills may be inaccurate due to recollection errors.
- The report of procedures deemed essential likely depends on supports available at a given location.
- Reports from those working at teaching centres may include supervision of and/or delegation to trainees, and thus the frequency of performing procedures may not be recounted accurately.



Conclusions

- Canadian paediatricians perform a wide range of procedural skills with varying frequency.
- Procedures that are performed with high frequency or deemed essential to independent practice (such as infant bag-valve mask ventilation and lumbar puncture) should be priorities for post-graduate training.
- Further analyses investigating for associations of procedural skills with geographic location and practice type should be conducted.



Anticipated survey impact

- The survey results represent the most comprehensive procedural skills needs assessment for Canadian general paediatricians. Results will be used to inform post-graduate education curricula, particularly as training programs transition to competency-based medical education.
- The results will also be used to highlight procedural skills that should be considered priorities for continuing professional training.



Publication and dissemination

Procedural skills needs for Canadian paediatricians: A national snapshot. White J, Gupta R, Writer H, Rowan-Legg A. Canadian Paediatric Society Annual Conference, Toronto, in June 2019 (oral presentation)

Demystifying defibrillators: A practical review for paediatricians. Gupta R, Writer H, White J. Canadian Paediatric Society Annual Conference, Toronto, in June 2019 (workshop)

Teething necklaces and bracelets worn by infants and toddlers

January 2018



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? Question

- Over the preceding 12 months, how often did paediatricians observe cases of serious adverse events related to the use of teething necklaces and bracelets?
- Are paediatricians encountering these products in their practice settings, and are they counselling families on the risks associated with their use?

! Importance

Necklaces and bracelets made of amber or hazel wood beads have been widely promoted as a “natural” method of teething pain relief in infants and toddlers, despite the lack of scientific evidence to support their effectiveness. Such products pose significant risks such as choking, strangulation, and even death.

↻ Methodology

A one-time survey was sent to paediatricians and paediatric subspecialists through the Canadian Paediatric Surveillance Program (CPSP). The survey tool can be accessed at www.cpsp.cps.ca/surveillance.

✓ Results

The survey response rate was 36% (1,020/2,845).

Adverse events

- Survey respondents reported 10 cases of adverse events related to the use of teething necklaces and bracelets over the past 12 months, excluding duplicates (based on the first three digits of the postal code).
- Adverse events included swallowing the clasp magnet, strangulation by the necklace, choking on the beads, and contact dermatitis.

Treatment and outcomes

- These adverse events resulted in hospitalization, and in one case, an invasive medical procedure.
- None of the adverse events resulted in disability or death.

Paediatricians' awareness and experience

- Of the paediatricians who answered the survey, 77% (779/1,012) were aware that amber, hazel wood, or other teething necklaces were marketed to parents as a “natural” method to relieve teething symptoms.
- Ninety-one percent (918/1,012) of respondents were aware of the risks associated with teething necklaces and bracelets.
- Sixty-nine percent (653/950) of respondents reported having seen an infant or a toddler in their practice wearing a teething necklace or bracelet over the past 12 months.
- Only 13% (124/948) of responding paediatricians said they had been approached by families with questions regarding the use of teething necklaces and bracelets over the past 12 months.
- Fifty-nine percent (529/901) of respondents reported discussing the risks associated with teething necklaces and bracelets with families who were using the product. Only 8% (67/889) reported discussing these products with all families.

Survey limitations

- Given the nature of the survey, the total incidence of teething necklace and bracelet use was not captured.
- The survey likely captured an under-estimate of serious adverse events associated with these products, noting that this survey engaged only a subset of practising paediatricians, and did not capture the experience of family physicians.



Conclusions

- Life-threatening events associated with the use of teething necklaces and bracelets are occurring in Canada.
- Paediatricians are seeing infants and toddlers in their practices wearing teething necklaces or bracelets but not all are discussing the risks with families using these products.
- Paediatricians and other health care workers should advise families of the dangers associated with teething products, including education about the potential risks associated with their use, as a component of anticipatory guidance during routine health visits in infancy.
- Providing parents with advice on safe teething remedies, such as those recommended by the Canadian Paediatric Society (CPS), is strongly recommended.



Anticipated survey impact

Findings of this CPSP survey will be shared with Health Canada and may be used to inform work underway to regulate teething necklaces and bracelets to reduce/eliminate potential hazards.



Publication and dissemination

Teething necklaces and bracelets pose significant danger to infants and toddlers. Abdulsatar F, Matsui D, Miller M, Taheri S. *Paediatr Child Health* 2018 Nov 7; pxy155, <https://doi.org/10.1093/pch/pxy155> [Epub ahead of print]

Teething necklaces and bracelets pose significant danger to infants and toddlers. Abdulsatar F. Canadian Paediatric Society Annual Conference, Toronto, in June 2019 (poster presentation)

Publications 2016–2018

Published papers related to studies and one-time surveys

(For a complete list with hyperlinks, see www.cpsp.cps.ca/publications/published-papers-related-to-studies.)

Adrenal suppression

Symptomatic adrenal suppression among children in Canada. Goldbloom EB, Mokashi A, Cummings EA, Abish S, Benseler SM, Huynh H, Watson W, Ahmet A. *Arch Dis Child* 2017 Apr;102(4):338–9

All-terrain vehicle safety

All-terrain vehicle serious injuries and death in children and youth: A national survey of Canadian paediatricians. Gill PJ, McLaughlin T, Rosenfield D, Moore Hepburn C, Yanchar NL, Beno S. *Paediatr Child Health* 2018 Jun 18; pxy059, <https://doi.org/10.1093/pch/pxy059> [Epub ahead of print]

Early-onset eating disorders

From questions to answers: Examining the role of pediatric surveillance units in eating disorder research. Katzman DK, Madden S, Nicholls D, Mawjee K, Norris ML. *Int J Eat Disord* 2017 Mar;50(3):259–65

Early-onset neonatal sepsis

Population-based study of early-onset neonatal sepsis in Canada. Sgro M, Kobylanskii A, Yudin MH, Tran D, Diamandakos J, Sgro J, Campbell DM. *Paediatr Child Health* 2018 Apr 24; pxy018, <https://doi.org/10.1093/pch/pxy018> [Epub ahead of print]

E-cigarettes

E-cigarettes: A new hazard for children and adolescents. Richmond SA, Pike I, Maguire JL, Macpherson A. *Paediatr Child Health*, 2018; 23(4):255–9

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Major depressive disorder among pre-adolescent Canadian children: Rare disorder or rarely detected? Korczak DJ, Ofner M, LeBlanc J, Wong S, Feldman M, Parkin PC. *Acad Pediatr* 2017;17(2):191–7

Neonatal hyperbilirubinemia

Severe neonatal hyperbilirubinemia decreased after the 2007 Canadian guidelines. Sgro M, Kandasamy S, Shah V, Ofner M, Campbell D. *J Pediatr* 2016 Apr;171:43–7

Persistent albuminuria

Persistent albuminuria in children with type 2 diabetes: A Canadian Paediatric Surveillance Program study. Sellers EA, Hadjiyannakis S, Amed S, Dart AB, Dyck RF, Hamilton J, Langlois V, Panagiotopoulos C, Dean HJ. *J Pediatr* 2016 Jan;168:112–7

Severe iron-deficiency anemia

Severe iron-deficiency anaemia and feeding practices in young children. Parkin PC, DeGroot J, Maguire JL, Birken CS, Zlotkin S. *Public Health Nutr* 2016;19(4):716–22

CPSP Highlights published in *Paediatrics & Child Health*

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Rh sensitization

Rh sensitization in Canada is not obsolete. Baker JM, Campbell DM, Bhutani VK, Sgro M. *Paediatr Child Health* 2017;22(4):238–9

Self-harm

Serious self-harm requiring intensive care unit admission: Understanding near-fatal suicide attempts. Korczak DJ, Skinner R, Dopko R. *Paediatr Child Health* 2018 Jul 25; pxy077, <https://doi.org/10.1093/pch/pxy077> [Epub ahead of print]

Sentinel injuries

Minor injuries... major implications: Barrett R, Ornstein A, Hanes L. Watching out for sentinel injuries. *Paediatr Child Health* 2016;21(1):29–30

Severe microcephaly

Small bite, big problem: Understanding severe microcephaly in Canada. Nelson CRM, Demarsh A, Miller SP, Morris SK, Moore Hepburn C, Bitnun A, Moore A, Shevell M, Evans J, Tataryn J. *Paediatr Child Health* 2017; 22(8):504–5

Severe vitamin D deficiency

Severe vitamin D deficiency: A persistent yet preventable problem among Canadian youth. Ward LM, Ladhani M, Zlotkin S. *Paediatr Child Health* 2017;22(1):43–4

Teething necklaces

Teething necklaces and bracelets pose significant danger to infants and toddlers. Abdulsatar F, Matsui D, Miller M, Taheri S. *Paediatr Child Health* 2018 Nov 7; pxy155, <https://doi.org/10.1093/pch/pxy155> [Epub ahead of print]

Presentations in 2018

(For a complete list with hyperlinks, see www.cpsp.cps.ca/publications/presentations.)

National

All-terrain vehicle safety

All-terrain vehicle serious injuries and death in children and youth: A national survey of Canadian paediatricians. Gill P, McLaughlin T, Rosenfield D, Moore-Hepburn C, Yanchar N, Beno S. Trauma Association of Canada Annual Scientific Meeting and Conference, Toronto, in February (poster)

Cannabis

Canadian paediatricians' views and knowledge about cannabis use for medical purposes among children and adolescents. Bélanger RE, Grant C, Côté M, Donner E, Breakey V, Laflamme J, Pinard A-M, Rieder M. Canadian Paediatric Society Annual Conference, Quebec City, in May (oral and poster)

How to address cannabis use with teenagers and their parents in this era of legalization. Bélanger R, Grant C. Canadian Paediatric Society Annual Conference, Quebec City, in May (oral)

Caring for children and youth from military families

Calling for a medical home for Canadian military families. Cramm H, Mahar A, MacLean C, Rowan-Legg A, Tam-Seto L. 2018 Family Medicine Forum Conference, Toronto, in November (oral)

Current knowledge and needs of Canadian paediatricians delivering healthcare to children and youth in military families. Cramm H, Mahar A, MacLean C, Rowan-Legg A, Tam-Seto L. Canadian Association of Health Services and Policy Research Conference, Montreal, in May (oral)

Non-type 1 diabetes

Non-type 1 diabetes. Amed S, Sellers E. Canadian Paediatric Society Annual Conference, Quebec City, in May (oral)

Rh sensitization

Rh sensitization: A two-year Canadian national surveillance study. Baker JM, Campbell D, Pavenski K, Decou ML, Hollamby K, Jegathesan T, Bhutani V, Zipursky A, Sgro M. The Hospital for Sick Children Haematology Research Day, Toronto, in November (oral)

Self-harm

Suicide risk among children and adolescents: Making evidence-informed decisions. Korczak D. Canadian Paediatric Society Annual Conference, Quebec City, in May (oral)

Severe microcephaly

Severe microcephaly in Canada. Nelson C. Canadian Paediatric Society Annual Conference, Quebec City, in May (poster)

Tuberculosis

Paediatric tuberculosis in Canada: 2018 update and focus on latent infection. Kitai I. Canadian Paediatric Society Annual Conference, Quebec City, in May (oral)

Final results of national surveillance of childhood tuberculosis in Canada: 2013–2016. Giroux R. Canadian Paediatric Society Annual Conference, Quebec City, in May 2018 (poster)

International**Listeria in the newborn**

Listeriosis in neonates and infants in Switzerland and Canada. Huang S, Abu Raya B, Jost M, Bortolussi R, Bettinger J, Grabowski J, Lacaze T, Robinson J, Posfay Barbe K, Mäusezahl M, Kollmann TR. International Network of Paediatric Surveillance Units 10th Scientific Conference, Glasgow, Scotland, in March (oral)

Medical Assistance in Dying

Medical Assistance in Dying (MAID) a Canadian perspective. Moore Hepburn C. International Network of Paediatric Surveillance Units 10th Scientific Conference, Glasgow, Scotland, in March (oral)

Hypoglycemia

Monitoring for hypoglycaemic newborns - should we expand our risk categories? Flavin M, Osiovič H, Coughlin K, Gregoire K, Hu L, León JA, Ray J, Sgro M, Barr L, Grewal K, Gallipoli A. International Network of Paediatric Surveillance Units 10th Scientific Conference, Glasgow, Scotland, in March (oral)



Canadian Paediatric Surveillance Program

New Study and One-Time Survey Opportunities

The opportunity

- Benefit from the CPSP's well-established, timely, cost-effective, and internationally recognized surveillance platform.
- The CPSP is effective at monitoring low-frequency, high-impact diseases and conditions encountered by general paediatricians and paediatric subspecialists.

Track record

- The average monthly response rate from approximately 2,800 paediatricians is 80%.
- The average detailed questionnaire response rate varies between 80% to 90%.

Themes of interest

Including examples of successful CPSP studies

- Rare diseases (including genetic, metabolic, or rare acquired conditions)
 - Congenital myotonic dystrophy
 - Medium-chain acyl-coenzyme A dehydrogenase deficiency
- Rare complications of more common diseases
 - Adrenal suppression with glucocorticoid therapy
 - Serious adverse events associated with complementary and alternative medicine
- Emerging infections
 - Congenital Zika syndrome
 - Lyme disease
- Threats to public health and safety
 - Neonatal abstinence syndrome
 - Teething necklaces and bracelets worn by infants and toddlers

Study success factors

- A study or condition with an incidence of less than 500 cases per year
- A multidisciplinary study team, with national representation
- Local champions who encourage study reporting at their institutions

Study impact

Knowledge translation: Studies have been published in high-impact, peer-reviewed journals; the CPSP is well known and recognized by prominent editorial boards.

Public health policies and legislation: Results have informed the total ban on baby walkers and the promotion of booster seats to prevent lap-belt syndrome.

Professional medical guidelines: Results have informed guidelines such as the Canadian Paediatric Society position statements on neonatal hyperbilirubinemia and medical assistance in dying.

Public health promotion and education: Results have informed efforts to prevent vitamin D deficiency rickets and the use of e-cigarettes in those under the legal age to use conventional tobacco products.

"As the Paediatric Chairs of Canada representative to the CPSP Steering Committee, I have witnessed the extraordinary ability of the CPSP to bring together study investigators from across paediatric disciplines and across Canada in the study of rare paediatric diseases. For conditions that are high in disability, morbidity, mortality, and economic costs to society, despite their low frequency, national surveillance to capture case-level data is essential. On behalf of the Steering Committee I would like to extend a sincere thank you to the thousands of CPSP participants who contribute to the Program. We are truly fortunate to have such a robust paediatric surveillance program in Canada."

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