

# 2014 Results

CANADIAN PAEDIATRIC SURVEILLANCE PROGRAM





## **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

Genetics

**Infectious  
diseases**

Injury  
prevention

Mental  
health

Neurology

Both vaccine-preventable diseases (VPDs) and non-VPDs continue to be a global public health challenge. Until these diseases are eradicated worldwide, Canadian children remain at risk, and surveillance is essential.

## Surveillance of infectious diseases a priority

### How the CPSP contributes

- ▶ **Informs** policy-makers about VPDs in Canada: congenital rubella syndrome, necrotizing fasciitis, subacute sclerosing panencephalitis, and acute flaccid paralysis (polio)
- ▶ **Identifies** clinical information on risk factors, management and outcomes for reportable infectious illnesses, including childhood tuberculosis, MRSA, and hemolytic uremic syndrome
- ▶ **Mobilizes** public health systems and prevention strategies around non-VPDs: congenital CMV infection, early-onset neonatal sepsis and meningitis, RSV infections in transplant patients, and neonatal herpes simplex virus infection
- ▶ **Develops** targeted professional and public education initiatives to inform those caring for children and youth about VPDs and non-VPDs: pre-travel anticipatory guidance and congenital CMV diagnosis and treatment

Successful surveillance requires complete and accurate reporting of cases to identify disease trends, clinical characteristics, and outcomes. The CPSP is grateful for the support of its participants.



[www.cpsp.cps.ca](http://www.cpsp.cps.ca)

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# Tribute to Dr. Danielle Grenier



The Canadian Paediatric Surveillance Program (CPSP) would like to pay special tribute to one of its founding members, Dr. Danielle Grenier, who passed away in September 2014.

Dr. Grenier's enthusiasm and unwavering belief that active national surveillance on rare childhood diseases or conditions made a true difference was felt by everyone who had the opportunity to cross paths with her over the years.

She strongly urged all paediatricians and paediatric subspecialists in Canada to participate in the CPSP either through monthly reporting or as investigators. She was committed to ensuring that program results be used to inform policies, guidelines, education, and advocacy for children and youth.

Alongside her commitment to Canadian children and youth, she was engaged at the international level, and encouraged countries to conduct simultaneous surveillance studies where results could be compared and lead to universal recommendations. She was highly effective in ensuring that actions were principle-driven and child-focused at all times.

Among her many other accomplishments, Dr. Grenier held the position of the Canadian Paediatric Society Medical Affairs Director from 1994 to 2014, had nearly 40 articles to her credit, and was a true inspiration to paediatric residents, colleagues, staff, and friends. All of this was in addition to being a full-time community paediatrician known for her exemplary care, compassion, and good humour. She was cherished by her husband, fellow paediatrician Dr. Luc Charette, and her sons Philippe and Laurent.

We are indebted to Dr. Grenier for all she did for children and youth, both in Canada and around the world.

# Foreword

## President of the Canadian Paediatric Society

### Dr. Robert Moriartey

As President of the Canadian Paediatric Society and a community paediatrician practising in Alberta for 36 years, I am proud to participate in the Canadian Paediatric Surveillance Program.



The CPSP is an important vehicle for both general paediatricians and subspecialists. This active surveillance network allows us to obtain additional information on diseases and conditions we may only encounter once or twice during our entire careers. By participating in the program, we can better understand these conditions and continue to offer our paediatric patients effective management and treatment options.

I invite all of my fellow paediatricians to take a few moments to discuss with your colleagues the opportunity to submit new study ideas to the CPSP. The CPSP Steering Committee always welcomes meeting with research teams and helping them develop successful studies that span a wide range of subjects and age groups. Over the years, many of the study and survey results have informed policies and position statements, provided evidence to advocate for the ban of unsafe products, identified potential adverse events to medications, and even alerted us to potential health risks due to events occurring in other countries.

Thanks to all the CPSP participants who contribute to the program on a regular basis – your involvement truly makes a difference!

## CPSP Chair

### Dr. Kimberly Dow

In 2014, the Canadian Paediatric Surveillance Program's Steering Committee took the opportunity to reflect on the program's achievements and discuss its vision for the next few years. Committee members identified six key priority areas: maintaining a strong partnership with the Public Health Agency of Canada, increasing knowledge translation and publications on study and survey results, continuing to engage strong research teams, maintaining active involvement from CPSP participants, using study results to inform advocacy efforts, and finally, linking with external databases to validate study results. Work on these various outcomes will take place in 2015 and in future years.



In 2014, the program was proud to launch three new studies: sudden unexpected death in epilepsy, childhood Lyme disease, and hypoglycemia in low-risk term newborns.

Thank you to all the CPSP study investigators who have worked diligently to analyse the results and further our knowledge of the risk factors, management, and prevention of many rare paediatric conditions.

As CPSP Chair, I want to take this opportunity to thank all of the participants who faithfully completed their monthly report forms in 2014 and also took the time to complete detailed questionnaires on the various diseases and conditions. Without your support and commitment, the CPSP could not exist.

# 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 2014 from the Public Health Agency of Canada, Health Canada's Therapeutic Effectiveness and Policy Bureau, and the following non-governmental sources:

- Citizens United for Research in Epilepsy
- Novartis Pharmaceuticals Canada Inc.
- Ontario Brain Institute
- Queen's Pediatrics Departmental Development and Innovation Fund
- SickKids Foundation

# CPSP Steering Committee

Kimberly Dow, MD (Chair)	Canadian Paediatric Society
Peter Buck, DVM, MSc	Centre for Food-borne, Environmental and Zoonotic Infectious Diseases, Public Health Agency of Canada
Claude Cyr, MD	Canadian Paediatric Society
Marie Adèle Davis, MBA	Canadian Paediatric Society
Elizabeth Donner, MD	Canadian Association of Child Neurology (Liaison)
Ciaran Duffy, MB	Paediatric Chairs of Canada
Kevin Gordon, MD	Canadian Association of Child Neurology (Liaison)
Danielle Grenier, MD	Canadian Paediatric Society
Margaret Herbert, MSc	Centre for Chronic Disease Prevention, Public Health Agency of Canada
Krista Jangaard, MD	Canadian Paediatric Society
W. James King, MD	Canadian Paediatric Society
Melanie Laffin Thibodeau, BCom	Canadian Paediatric Society
Jonathon Maguire, MD	Canadian Paediatric Society
Dorothy Moore, MD	IMPACT (Immunization Monitoring Program ACTive) (Liaison)
Paul Muirhead, LL.M.	Consultant
Jorge Pinzon, MD	Canadian Paediatric Society
Anne Rowan-Legg, MD	Canadian Paediatric Society
Dorcas Taylor, MSc	Centre for Communicable Diseases and Infection control, Public Health Agency of Canada
Paul Thiessen, MD	Canadian Paediatric Society
Lesley Ann Turner, MD	Canadian College of Medical Geneticists (Liaison)

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In April 2014, **Kevin Gordon** completed a nine-year term as the liaison for the Canadian Association of Child Neurology on the CPSP Steering Committee, **Lesley Ann Turner** completed a six-year term as the liaison for the Canadian College of Medical Geneticists and **Paul Thiessen** completed a four-year term as a Canadian Paediatric Society representative. We sincerely thank them for their dedication to the program and expertise on the committee. We wish them all the best in future endeavours.

The committee also wishes to thank **Anne-Marie Ugnat** from the Public Health Agency of Canada (PHAC) who provided seven years of leadership and expertise as the PHAC liaison on the CPSP Working Group and Steering Committee. We wish Anne-Marie every success in future projects and in her new role at PHAC.

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# CPSP Working Group

Melanie Laffin Thibodeau, BCom (Chair)	Canadian Paediatric Society
Marie Adèle Davis, MBA	Canadian Paediatric Society
Danielle Grenier, MD	Canadian Paediatric Society
Margaret Herbert, MSc	Centre for Chronic Disease Prevention, Public Health Agency of Canada
Melanie Khalil, BA	Canadian Paediatric Society
Jonathon Maguire, MD	Canadian Paediatric Society
Anne-Marie Ugnat, PhD	Centre for Chronic Disease Prevention, Public Health Agency of Canada



# Publications 2010–2014

## 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](http://www.cpsp.cps.ca/publications/published-papers-related-to-studies).)

### Acute flaccid paralysis

Performance of acute flaccid paralysis surveillance compared to WHO standards. Desai S, Smith T, Thorley BR, Grenier D, Dickson N, Altpeter E; SPSU Committee, Sabbe M, Elliott E, Zurynski Y. *J Paediatr Child Health* 2014 Jul 30. doi: 10.1111/jpc.12691. [Epub ahead of print]

### Adverse events following immunization

Canadian paediatricians' approaches to managing patients with adverse events following immunization: The role of the Special Immunization Clinic network. Top KA, Zafack J, De Serres G, Halperin SA. *Paediatr Child Health* 2014;19(6):310–4

### Child maltreatment

Head injury secondary to suspected child maltreatment: Results of a Canadian national surveillance program. Bennett S, Ward M, Moreau K, Fortin G, King J, MacKay M, Plint A. *Child Abuse Negl* 2011;35(11):930–6

### Complementary and alternative medicine

Serious adverse events associated with pediatric complementary and alternative medicine. Zorzela L, Boon H, Mior S, Yager J, Gross A, Vohra S. *Eur J Integr Med* 2014;6(4):467–72

### Concussion management

Concussion management by paediatricians: A national survey of Canadian paediatricians. Gordon KE, Do MT, Thompson W, McFaul S. *Brain Inj* 2014;28(3):311–7

### Congenital cytomegalovirus infection

Congenital cytomegalovirus infection in Canada: Active surveillance for cases diagnosed by paediatricians. Vaudry W, Lee BE, Rosychuk RJ. *Paediatr Child Health* 2014;19(1):e1–5

### Congenital myotonic dystrophy

Congenital myotonic dystrophy in a national registry. Prendergast P, Magalhaes S, Campbell C. *Paediatr Child Health* 2010;15(8):514–8

Congenital myotonic dystrophy: Canadian population-based surveillance study. Campbell C, Levin S, Siu VM, Venance S, Jacob P. *J Pediatr* 2013;163(1):120–5.e1–3

### Congenital rubella syndrome

Rubella elimination: The Canadian experience. Macey JF, Tam T, Lipskie T, Tipples G, EisBrenner T. *J Infect Dis* 2011;204:S585–92

### Eating disorders

Incidence and age-specific presentation of restrictive eating disorders in children – A Canadian Paediatric Surveillance Program study. Pinhas L, Morris A, Crosby RD, Katzman DK. *Arch Pediatr Adolesc Med* 2011;165(10):895–9

### Food allergy

Canadian allergists' and nonallergists' perception of epinephrine use and vaccination of persons with egg allergy. Desjardins M, Clarke A, Alizadehfard R, Grenier D, Eisman H, Carr S, et al. *J Allergy Clin Immunol: In Practice* 2013;1:289–94

### **Kernicterus / neonatal hyperbilirubinemia**

Incidence of chronic bilirubin encephalopathy in Canada, 2007–2008. Sgro M, Campbell DM, Kandasamy S, Shah V. *Pediatrics* 2012;130(4):e886–90

Acute neurological findings in a national cohort of neonates with severe neonatal hyperbilirubinemia. Sgro M, Campbell D, Barozzino T, Shah V. *J Perinatol* 2011;31(6):392–6

### **Medium-chain acyl-CoA dehydrogenase deficiency**

Incidence of medium-chain acyl-CoA dehydrogenase deficiency in Canada using the Canadian Paediatric Surveillance Program: Role of newborn screening. Prasad C, Speechly KN, Dyak S, Rupar CA, Chakraborty P, Kronick JB. *Paediatr Child Health* 2012;17(4):185–9

### **Methicillin-resistant *Staphylococcus aureus* (MRSA)**

Clinical characteristics of pediatric patients hospitalized with methicillin-resistant *Staphylococcus aureus* in Canadian hospitals from 2008 to 2010. Leifso KR, Gravel D, Mouchili A, Kaldas S, Le Saux N. *Can J Infect Dis Med Microbiol* 2013;24(3):e53–6

### **Non-type 1 diabetes mellitus**

Differing clinical features in Aboriginal vs. non-Aboriginal children presenting with type 2 diabetes. Amed S, Hamilton JK, Sellers EAC, Panagiotopoulos C, Hadjiyannakis S, Shah BR, Booth GL, Laubscher TA, Dannenbaum D, Dean H. *Pediatr Diabetes* 2012;13(6):470–5

Risk factors for medication-induced diabetes and type 2 diabetes. Amed S, Dean H, Sellers EAC, Panagiotopoulos C, Shah BR, Booth GL, et al. *J Pediatr* 2011;159(2):291–6

Type 2 diabetes, medication-induced diabetes, and monogenic diabetes in Canadian children: A prospective national surveillance study. Amed S, Dean HJ, Panagiotopoulos C, Sellers EAC, Hadjiyanakis S, Laubscher TA, Dannenbaum D, Shah BR, Booth GL, Hamilton JK. *Diabetes Care* 2010;33(4):786–91

### **Paediatric myasthenia**

Clinical characteristics of paediatric myasthenia: A surveillance study. VanderPluym J, Vajsar J, Jacob FD, Mah JK, Grenier D, Kolski H. *Pediatrics* 2013;132(4):e939–44

### **Severe combined immunodeficiency**

Severe combined immunodeficiency (SCID) in Canadian children: A national surveillance study. Rozmus J, Junker A, Thibodeau ML, Grenier D, Turvey SE, Yacoub W, Embree J, Haddad E, Langley JM, Ramsingh RM, Singh V-A, Long R, Schultz KR. *J Clin Immunol* 2013;33:1310–6

### **Transfusion-related acute lung injury**

Transfusion-related acute lung injury in the Canadian paediatric population. Gauvin F, Robillard P, Hume H, Grenier D, Whyte R, Webert KE, Fergusson D, Lau W, Froese N, Delage G. *Paediatr Child Health* 2012;17(5):235–9

## CPSP Highlights published in 2014 in *Paediatrics & Child Health*

(For a complete list with hyperlinks, see [www.cpsp.cps.ca/publications/cpsp-highlights](http://www.cpsp.cps.ca/publications/cpsp-highlights).)

Redefining feeding and eating disorders: What is avoidant/restrictive food intake disorder? Katzman DK, Stevens K, Norris M. *Paediatr Child Health* 2014;19(8):445–6

Sudden unexpected death in epilepsy: Who are the children at risk? Donner EJ. *Paediatr Child Health* 2014;19(7):389

Kawasaki disease: High index of suspicion needed in a febrile child. Scuccimarri R, Yeung RSM. *Paediatr Child Health* 2014;19(5):239–40

Paediatric methadone ingestions: An under-recognized form of child maltreatment? Lewington LE, Shaffer C, Ornstein AE. *Paediatr Child Health* 2014;19(3):139–40

Encounters with an old foe: Childhood tuberculosis in Canada. Kitai I, Morris SK. *Paediatr Child Health* 2014;19(2):89–90

# Presentations in 2014

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

## National

### Acute flaccid paralysis

Guillain-Barré syndrome after immunization in Canadian children (1996–2012). Top KA, Desai S, Moore D, Law BJ, Vaudry W, Halperin SA, Bettinger JA, for members of the Immunization Monitoring Program ACTIVE. Canadian Immunization Conference, Ottawa, in December (poster)

### Adrenal suppression

Adrenal suppression in the paediatric population in Canada. Goldbloom EB, Mokashi A, Cummings E, Huynh H, Benseler S, Watson W, Abish S, Ahmet A. Canadian Paediatric Society Annual Conference, Montréal, in June (oral)

### Eating disorders

An update on eating disorders and changes in the DSM-5. Katzman D, Norris M. Canadian Paediatric Society Annual Conference, Montréal, in June (oral)

### Hypoglycemia

Hypoglycemia in newborns with no pre-identified risk factors. Flavin M, Grewal K, Hu L. Canadian Paediatric Society Annual Conference, Montréal, in June (poster)

### Langerhans cell histiocytosis

Survey of Langerhans cell histiocytosis in Canada. Crooks B, Weitzman S, Dix D, Parker L. Histiocyte Society Annual Meeting, Toronto, in October (poster)

### Neonatal hyperbilirubinemia

Comparing rates of severe neonatal hyperbilirubinemia in three Western industrialized populations: Canada, the United Kingdom & Ireland, and Switzerland. Sgro M, Kandasamy S, Ofner M. PAS/ASPR Joint Meeting, Vancouver, in May (poster)

Surveillance of severe neonatal hyperbilirubinemia in Canada. Sgro MD, Kandasamy S, Campbell D, Ofner M, Shah V. PAS/ASPR Joint Meeting, Vancouver, in May (oral)

### Palliative care

Paediatric palliative care in Canada: A national survey of paediatricians. Cyr C, Maisonneuve M. Canadian Paediatric Society Annual Conference, Montréal, in June (poster)

## International

### Acute flaccid paralysis

Guillain-Barré syndrome after immunization in Canadian children (1996–2012). Top KA, Desai S, Moore D, Law BJ, Vaudry W, Halperin SA, Bettinger JA, for members of IMPACT. Vaccine and ISV Congress, Philadelphia, in October (poster)

### Conversion disorder

Conversion disorders in Canadian children and youth: A national survey of clinical features and treatment outcomes. Krasnik CE, Grant C. American Academy of Child and Adolescent Psychiatry Annual Meeting, San Diego, in October (poster)

### Periodic fever syndromes

Periodic fever syndromes in Canada: Results from a national surveillance program. Dancey P, Benseler S, Gattorno M, Junker AK, Laxer RM, Miettunen P, Turner LA. Pediatric Rheumatology Symposium, Orlando, in April (poster)

# Surveillance at Work

## Overview

The importance of surveillance to the practice of medicine cannot be overstated. Through the ongoing, systematic collection of data, the burden of disease can be determined, interventions to prevent the occurrence of a disorder can be assessed, and the information collected can be used to develop health policy. Surveillance takes research data into action.

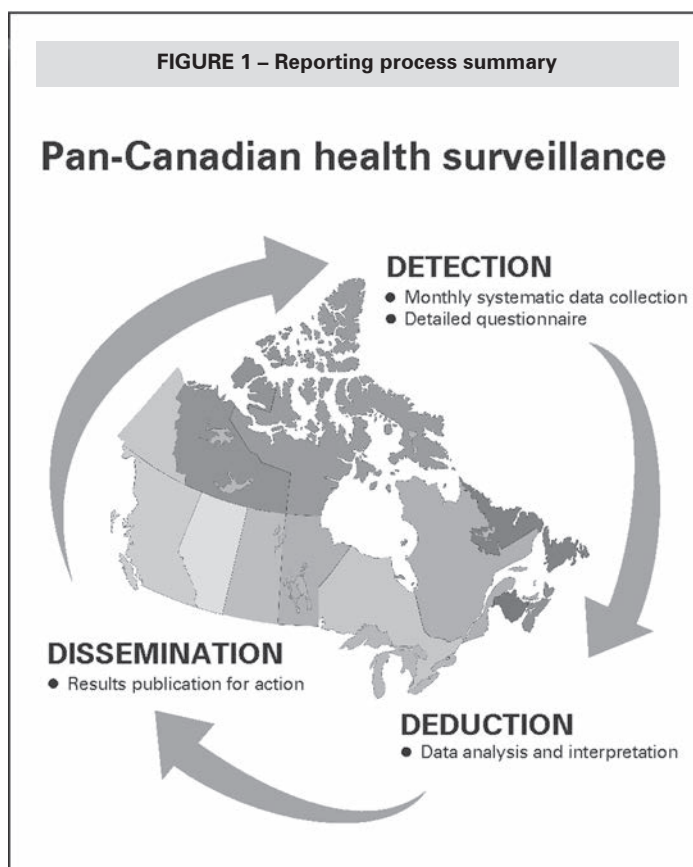
According to Statistics Canada, the Canadian population on July 1, 2014 was an estimated 35,540,419, with 7,847,367 individuals 0–19 years of age inclusively, which represents 22% of the population. Although individually uncommon, rare diseases affect hundreds of thousands of Canadian children and youth and typically have lifelong impacts. The incidence of many rare disorders is unknown, and yet is essential for improved clinical care, advocacy, and health service planning.

The CPSP provides an innovative means to undertake paediatric surveillance and increase awareness of childhood disorders that are high in disability, morbidity, mortality, and economic cost to society, despite their low frequency. Preference is given to studies that have strong scientific and public health importance or could not be undertaken any other way. All studies in the program must conform to high standards of scientific rigour and practicality, and the CPSP assures the confidentiality of all information collected. To enhance data capture on individual studies, the program works collaboratively with other professional groups, such as psychiatrists, pathologists/coroners, and adult infectious disease specialists.

## Process

The CPSP Steering Committee oversees the program and reviews new study proposals. Upon initiation of a new study, practising Canadian paediatricians, paediatric subspecialists, and other health care providers receive a summary of the protocol, including the case definition and a brief description of the condition. This serves to educate and increase awareness of the conditions under surveillance while providing a uniform basis for reporting. The CPSP uses a two-tiered reporting process to ascertain and investigate cases: an initial 'check-off' form and a detailed questionnaire. The full process is summarized in Figure 1 and includes the 3Ds of surveillance: detection, deduction, and dissemination.

Case ascertainment is undertaken by comparing a few selected study results with cases reported to the hospital Discharge Abstract Database of the Canadian Institute for Health Information (CIHI) and by investigating duplicate reports and comparing data with related programs or centres. To date, case ascertainment has been excellent.



## Reporting

The check-off form, listing the conditions currently under surveillance, is distributed monthly to participants. For each condition, respondents are asked to indicate the number of new cases seen in the last month, including 'nil' reports. A 'nil' report is very important in active surveillance; the CPSP cannot simply assume that no reply means there were no cases. In October 2011, the program launched eCPSP, an electronic platform giving participants the opportunity to receive their monthly forms online. By December 2014, 68% of program participants were signed up for electronic reporting.

Participants report all cases meeting the case definitions, including suspect or probable cases. This sometimes leads to duplicate reports but avoids missed cases. The list of studies conducted by the program since 1996 can be accessed at [www.cpsp.cps.ca/surveillance](http://www.cpsp.cps.ca/surveillance).

Confidentiality is maintained by using only non-nominal patient information, such as the date of birth and sex of the child. This anonymous information is used to exclude duplicates and is sent to the original respondent to request case-specific information.

Once the detailed questionnaire is returned to the CPSP, it is forwarded to the investigator for analysis. If further information is required to confirm or exclude a case, a member of the CPSP staff contacts the respondent on behalf of the investigator.

Participants who do not reply every month receive reminders. In addition, information on the monthly compliance rates and the number of cases reported is distributed quarterly to all participants to keep them informed of progress. In 2014, the national reporting rate was 80% (Table 1) and the response rate for completion of detailed questionnaires, 87% (Table 2).

**TABLE 1 – Initial response rates (%) and number of participants for 2014**

Provinces/territories	Reporting rates (%)	Number of participants
Alberta (AB)	79	356
British Columbia (BC)	77	257
Manitoba (MB)	79	119
New Brunswick (NB)	82	26
Newfoundland and Labrador (NL)	82	53
Nova Scotia (NS)	82	96
Northwest Territories (NT)	100	2
Nunavut (NU)	67	3
Ontario (ON)	80	980
Prince Edward Island (PE)	98	9
Quebec (QC)	77	547
Saskatchewan (SK)	77	56
Yukon (YT)	92	1
<b>Canada</b>	<b>80</b>	<b>2505</b>

**TABLE 2 – 2014 detailed questionnaire completion rates as of May 1, 2015**

Studies/conditions	Reported cases*	Pending	% Completion rate
Acute flaccid paralysis	55	7	87
Adverse drug reactions – serious and life-threatening	40	9	78
Childhood Lyme disease	23	4	83
Childhood tuberculosis	95	17	82
Early-onset major depressive disorder	14	2	86
Fragile X syndrome	6	0	100
Hypoglycemia in low-risk term newborns	63	16	75
Kawasaki disease	365	33	91
Periodic fever syndromes	47	5	89
Severe alcohol intoxication in adolescents	25	3	88
Sudden unexpected death in epilepsy	6	1	83
<b>Total number of cases (all studies)</b>	<b>739</b>	<b>97</b>	<b>87</b>

\* Excluding duplicate and excluded cases

## Participant workload

The monthly reporting system is simple and the follow-up study questionnaires are easy to complete. As only non-nominal, non-identifiable data are collected through the CPSP, respondents do not hesitate to provide clinical information.

In 2014, the majority of participants (86%) had 'nil' cases to report. The importance of zero reporting must be re-emphasized. Table 3 illustrates the number of cases reported by respondents in 2014. As studies come and go, the workload shifts to different subspecialties. Through the years, studies with national collaborative networks have been very successful. The 2014 study with the most reports was Kawasaki disease.

Number of cases	% Respondents
0	85.9
1–5	13.0
6–10	0.7
>10	0.4

The CPSP is extremely grateful that the majority of participants diligently complete the detailed questionnaires subsequent to reporting cases. This step suggests that they appreciate the enormous value of the scientific data collected and reinforces the Steering Committee's insistence on short, precise, and pertinent detailed questionnaires.

To thank paediatricians and paediatric subspecialists for their tremendous commitment and support, the names of participants who completed the initial reporting forms for all months in 2014 and/or returned one or more detailed questionnaires were entered in draws for various prizes.

## Investigators' corner

Through timely, active surveillance, the CPSP provides investigators with an innovative means of obtaining non-nominal, national data on low-frequency diseases and conditions from over 2500 participants. The program is committed to a high case-ascertainment rate and, due to follow-up reminders to non-respondents, obtained a response rate of 87% on detailed questionnaires (Table 2). The CPSP offers an opportunity for international collaboration with other paediatric surveillance units worldwide and a chance to make a difference in the health and well-being of Canadian children and youth.

Individual researchers are encouraged to submit proposals for new studies that meet the "criteria for inclusion," and to follow the "format for submission," available on the CPSP website at [www.cpsp.cps.ca/apply-proposez](http://www.cpsp.cps.ca/apply-proposez). The Steering Committee reviews submissions at its spring and fall meetings, giving preference to studies that have either strong scientific and public health importance or those that could not be undertaken any other way. Following the review, studies must receive ethical approval and have external funding arrangements in place before final acceptance to the program. Researchers interested in obtaining more information about the program are encouraged to visit the website, [www.cpsp.cps.ca](http://www.cpsp.cps.ca), or to contact the manager of surveillance at [cpssp@cpsp.ca](mailto:cpssp@cpsp.ca).

## One-time survey questions

The CPSP is also available to investigators as a cost-effective tool to survey participants on a one-time basis to identify the prevalence of a problem or answer a specific question. Once approved by the CPSP Steering Committee, the one-time survey is sent to all participants with the monthly initial reporting form. Collected results are forwarded to the investigator for data analysis.

Results of the 2014 one-time survey questions are found on pages 41–42, and the list of surveys completed to date can be accessed at [www.cpsp.cps.ca/surveillance/one-time-surveys](http://www.cpsp.cps.ca/surveillance/one-time-surveys).

### International developments

The program 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 research. No other network enables international comparisons of demographics, diagnosis, treatments, and outcomes for rare childhood conditions.

Established in 1992, INoPSU now includes 12 paediatric surveillance units (PSUs) among its membership. Incredibly, many of the 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.



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#### Glossary of terms for tables of cases in each study results

Reported: Reports of cases received

Duplicates: Cases reported by more than one participant

Excluded: Cases not meeting the case definition

Pending: Detailed reports not received or not yet confirmed

Confirmed: Cases verified as meeting the case definition



# CPSP Principal Investigators

## Surveillance studies in 2014



Dr. Shalini Desai  
*Acute flaccid paralysis*



Margaret Zimmerman  
*Adverse drug reactions –  
serious and life-threatening*



Dr. Joanne Langley  
*Childhood Lyme disease*



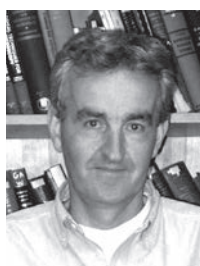
Dr. Ian Kitai  
*Childhood tuberculosis*



Dr. Daphne Korczak  
*Early-onset major  
depressive disorder*



Dr. Gudrun Aubertin  
*Fragile X syndrome*



Dr. Michael Flavin  
*Hypoglycemia in low-risk  
term newborns*



Dr. Rosie Scuccimarrì  
*Kawasaki disease*



Dr. Paul Dancey  
*Periodic fever syndromes*



Dr. Amy Acker  
*Severe alcohol  
intoxication in  
adolescents*



Dr. Elizabeth Donner  
*Sudden unexpected  
death in epilepsy*

# Surveillance Studies in 2014

## Acute flaccid paralysis

Ongoing study since January 1996

J Rotondo, R Pless, S Desai, T Smith, S Squires

### Highlights 2014

- In accordance with World Health Organization (WHO) recommendations, Canada conducts acute flaccid paralysis (AFP) surveillance in the under 15 population to monitor for polio in light of ongoing transmission of wild poliovirus in countries around the world.
- Thirty-seven cases of AFP were confirmed in 2014, representing a non-polio AFP detection rate of 0.65 cases per 100,000 children less than 15 years of age, well below the rate of 1.0 case per 100,000 population required to maintain Canada's status as a polio-free region.
- All AFP cases were thoroughly investigated and none were diagnosed as poliomyelitis. The most common diagnoses were Guillain-Barré syndrome (GBS) and transverse myelitis (TM).
- The AFP surveillance detailed questionnaire has been updated and a user manual developed with the aim of improving Canada's ability to meet the WHO surveillance performance objectives.

### Background and objectives

The complete protocol (updated in 2015) can be accessed at [www.cpsp.cps.ca/surveillance](http://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.

### Results

In 2014, 55 reports of AFP were made to the Public Health Agency of Canada (PHAC), 29 (53%) from the CPSP network and 26 (47%) from the Canadian Immunization Monitoring Program ACTive (IMPACT). Eighteen cases were excluded from the analysis: 8 were duplicates, 2 did not meet the age criteria,

1 was lost to follow-up, and 7 had detailed questionnaires still pending. Thirty-seven cases met the national case definition. No cases of poliomyelitis were identified. The 37 confirmed cases represent a non-polio AFP detection rate of 0.65 cases per 100,000 children less than 15 years of age (Table 2). Of the confirmed cases, the average time from case onset to reporting to CPSP was 109 days (range: 13 to 458).

Reported	Duplicates	Excluded	Pending	Confirmed
55	8	3	7	37

Cases ranged in age from <1 to 14 years with a mean of 7.2 years (95% CI 5.7–8.6) and a median of 6.5 years. These numbers are consistent with those of previous years (mean: 6.8 [95% CI 6.5–7.1], median: 5.9). Twenty (54%) cases were male and 17 (46%) were female, which is also consistent with the gender distribution observed in previous years (59% male, 41% female).

Documentation of age-appropriate polio immunization was provided for 18 (49%) cases, 3 (8%) were recorded as “up-to-date” with their immunizations (no further information was available), 3 (8%) were not up-to-date for their immunizations, and the remaining 13 (35%) did not have information regarding immunization. Poliovirus vaccine coverage in Canada is estimated to be 96% for three or more doses among 2-year-olds, 96% for four or more doses among 7-year-olds, and 95% for four or more doses among 17-year-olds.

### Medical history and clinical features

Among the 37 cases, 1 (3%) was considered immunocompromised and 3 (8%) had an abnormal neurological history (febrile seizure, developmental delay, and trauma). Eleven (30%) cases had a history of acute respiratory illness within the 30 days prior to AFP onset, of which 6 had a viral etiology identified as causing the respiratory illness: 2 cases had enterovirus D68 (EV D68), 1 had either an enterovirus or rhinovirus, 1 had influenza A, and 2 had influenza B.

Ten (27%) cases reported experiencing fever at the onset of paralysis. All but one case had information regarding the distribution of weakness, with 5 (14%) cases experiencing unilateral weakness, 30 (81%) experiencing bilateral

weakness, and 1 case having only the cranial nerves affected. Three (8%) cases experienced weakness in one limb, 8 (22%) in two limbs, 2 (5%) in three limbs, and 22 (60%) in four limbs. Seven (19%) cases had their respiratory muscles affected and 2 (5%) had cranial nerves affected. The average time from symptom onset to greatest weakness was 10 days (median: 7 days, range: <1 to 60).

### Investigation for enteroviruses and *Campylobacter*

Four (11%) cases had no samples obtained for virological investigations, 19 (51%) had samples obtained from one source, and 14 (38%) had samples obtained from more than one source. Sources included stool for 7 (19%) cases, cerebrospinal fluid (CSF) for 24 (65%) cases, throat swabs for 11 (30%) cases, nasopharyngeal swabs for 9 (24%) cases, and a blood sample for 1 (3%) case. Where stool was collected, 3 (43%) were considered adequate (i.e., taken within 14 days of the paralysis onset). Stool collection for the remaining 4 (57%) cases occurred later, when the sensitivity of enterovirus isolation would have been lower. None of the samples were positive for polioviruses. Other viral etiologies identified include adenovirus or parainfluenza virus in 2 cases, enterovirus in 3 cases (2 of which were EV D68), and rhinovirus in 2 cases. Testing conducted for *Campylobacter* in 7 cases yielded negative results for all but 1 case whose species was *Campylobacter jejuni*.

### Neurological investigations and final diagnoses

All cases underwent at least one type of neurological investigation, the most frequent being CSF examination (35 cases, 95%) and MRI/CT scans (33 cases, 89%). Among tested cases, 21/35 (60%) had abnormal CSF chemistry results, 14/18 (78%) had abnormal electromyography and/or nerve conduction studies, and 25/33 (76%) had an abnormal MRI or CT scan. As observed in previous years, the majority of AFP cases (21 cases, 57%) were diagnosed as GBS, 2 of which were a Miller-Fisher variant and 1 was acute motor axonal neuropathy. Ten (27%) cases were diagnosed as TM. The remaining cases were diagnosed as follows: acute disseminated encephalomyelitis (1 case), AFP and acute monoparesis secondary to EV D68 infection (2 cases), Bickerstaff encephalitis secondary to *Campylobacter jejuni* infection (1 case), chronic inflammatory demyelinating polyneuropathy (1 case), and “post-infectious” AFP (1 case).

### Hospitalization and outcomes

Every case was hospitalized; length of stay ranged from 1 to 120 days with a mean of 13.9 days (95% CI 6.9–20.9) and a median of 8.0 days. This was consistent with previous years (mean: 13.5 [95% CI 12.3–14.7], median: 8.0). Outcome at the time of the initial report was documented in 32 (87%) of the cases: 3 (9%) fully recovered, 26 (81%) partially recovered with residual weakness, and 3 (9%) had not recovered. Thirteen (35%) cases had clinical outcomes reported at 60 days, including 4 cases who had fully recovered, 7 who partially recovered, and 2 who had not recovered.

## Discussion

Although Canada and the rest of the Americas was certified as polio-free in September 1994 by WHO, there is an ever-present risk of poliovirus importation and spread within Canada so long as transmission is still occurring in other regions of the world. In 2014, polio remained endemic in only three countries worldwide (Pakistan, Afghanistan, and Nigeria). Nonetheless, numerous poliovirus exportations to polio-free countries and the threat these posed to the Global Eradication Initiative’s aim to eradicate polio by 2018, led WHO’s Director-General to declare the international spread of wild poliovirus in 2014 a public health emergency of international concern under the 2005 International Health Regulations. This event served as a reminder of the importance of continued vigilance in countries where polio has long been eliminated. Canada’s AFP surveillance system remains a core program that assists in monitoring for polio, provides a baseline for AFP incidence and causes in Canada, and maintains Canada’s polio-free status with WHO.

**TABLE 2 – Measure of WHO AFP surveillance targets in Canada, 1996–2014**

Year	Number of cases	Incidence rate*	% with adequate stool sample†	% with 60 days follow-up‡
1996	27	0.45	18.5	70.4
1997	35	0.59	28.6	45.7
1998	43	0.72	25.6	46.5
1999	60	<b>1.01</b>	33.3	33.3
2000	64	<b>1.09</b>	45.3	37.5
2001	52	0.89	36.5	38.5
2002	44	0.75	31.8	34.1
2003	44	0.76	34.1	22.7
2004	38	0.66	44.7	28.9
2005	53	0.93	26.4	32.1
2006	39	0.69	20.5	43.6
2007	50	0.89	42.0	46.0
2008	42	0.75	33.3	42.9
2009	59	<b>1.05</b>	30.5	49.2
2010	47	0.84	34.0	55.3
2011	44	0.78	31.8	47.7
2012	33	0.58	45.5	36.4
2013	34	0.60	20.6	35.3
2014	37	0.65	8.1	35.1

\* Incidence rate per 100,000 population; numbers in bold font meet the WHO target of 1.0 case per 100,000 population less than 15 years of age.

† Adequate stool sample refers to one stool sample taken within 14 days of paralysis onset; WHO target: 80%

‡ WHO target: 80%.

## Surveillance Studies in 2014

Several previously identified quality assurance issues continue to be evident in 2014. As in previous years, Canada was unable to meet its current AFP surveillance system objectives – objectives that are based on performance indicators used by WHO. Historically, Canada has only met the non-polio AFP incidence target of 1.0 case per 100,000 population under 15 years of age three times since AFP surveillance began in 1996 (1999, 2000, and 2009) and has never met the targets of stool testing in 80% of cases and 60-day follow-up of 80% of cases. Although Canada's inability to meet the incidence target may be associated with lower baseline levels of non-polio AFP in Canada, under-reporting of cases most likely plays a strong role, particularly since AFP is not yet reportable in all provinces and territories, and adult neurologists who treat paediatric patients may not be part of the CPSP network. As this study is ongoing, delays in reporting may have occurred and numbers for 2014 are expected to change.

Canada's low rate of stool sampling is likely due to a low index of suspicion for poliovirus infection and rapid neurological investigations. The lack of 60-day follow-up information could stem from most AFP cases being discharged from acute care hospitals prior to the 60-day follow-up and the lack of access to patient records. Currently, direct follow-up with the patients and/or their physicians is not possible through CPSP.

In response to these concerns, several mitigation activities are being developed and implemented:

- **Revised detailed questionnaire:** PHAC, in collaboration with CPSP and IMPACT, has revised the AFP detailed questionnaire and developed a user manual. The revised documents clearly state the importance of stool testing, 60-day follow-up, and reporting cases to both CPSP and the local public health unit in jurisdictions where AFP is reportable.
- **Education:** An update to the 1997 protocol for the investigation of AFP and suspected paralytic poliomyelitis is currently in development and is expected to educate physicians in an era when many may not have ever treated a suspected case of poliomyelitis. Communication activities with the Canadian Public Health Laboratory Network and CPSP regarding the importance of submitting a stool sample to the National Microbiology Laboratory are also being implemented.
- **Electronic reporting:** The move to electronic reporting of AFP cases to CPSP and PHAC in the next few years is expected to result in increased AFP case reporting and a large reduction in reporting delays.

### Conclusion

- Although Canada did not meet the WHO performance indicators for national AFP surveillance in 2014, there was no evidence to suggest that any polio cases occurred in Canada in 2014 and Canada's polio-free status remains.
- The most common diagnoses of AFP in Canada in children less than 15 years of age were GBS and TM.

### Publications and presentations

Desai S, Smith T, Thorley BR, Grenier D, Dickson N, Altpeter E; SPSU Committee, Sabbe M, Elliott E, Zurynski Y. Performance of acute flaccid paralysis surveillance compared to WHO standards. *J Paediatr Child Health* 2014 Jul 30. doi: 10.1111/jpc.12691. [Epub ahead of print]

Top KA, Desai S, Moore D, Law BJ, Vaudry W, Halperin SA, Bettinger JA, for members of the Immunization Monitoring Program ACTive. Guillain-Barré Syndrome after Immunization in Canadian Children (1996-2012). Canadian Immunization Conference, Ottawa, in December. (Poster presentation)

Top KA, Desai S, Moore D, Law BJ, Vaudry W, Halperin SA, Bettinger JA, for members of IMPACT. Guillain-Barré syndrome after immunization in Canadian children (1996–2012). Vaccine and ISV Congress, Philadelphia, October 2014. (Poster presentation)

### References

Available upon request from the CPSP office

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## Adverse drug reactions – serious and life-threatening

Ongoing study since January 2004

M Zimmerman

### Highlights 2014

- In 2014, the study confirmed 28 paediatric adverse drug reaction (ADR) cases.
- Systemic antibacterials, antiepileptics, and cardiovascular drugs were the most frequently reported drug classes causing adverse reaction(s).
- The majority of ADR reports described skin and subcutaneous disorders, a finding consistently observed since surveillance began in 2004.

### Background and objectives

The complete protocol can be accessed at [www.cpsp.cps.ca/surveillance](http://www.cpsp.cps.ca/surveillance).

### Case definition

Serious and life-threatening adverse 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 health product, which occurs at any dose and results in emergency observation, hospitalization, persistent or significant disability or death

### Exclusions

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

### Results

From January 1 to December 31, 2014, 40 cases of suspected ADRs were reported and 28 cases were confirmed as meeting the case definition. Of the 28 confirmed cases, 16 were male (57%) and age ranged from 5 months to 17 years. The largest number of reports (13, 46%) involved children aged 6 to 12 years (Table 2).

All 28 confirmed cases were classified as serious, with more than one reason for seriousness reported in 7 cases. Two deaths were reported in 2014. Table 3 compares the reasons for seriousness over the last seven years. In 2014, outcome information for all 28 cases was as follows: fatal (2, 7%); recovered (20, 71%); recovering/resolving (1, 4%); not yet recovered (3, 11%); and unknown (2, 7%).

The majority of the reports described skin and subcutaneous disorders that are documented in the Canadian approved product monograph. When an approved product monograph was not available, the information source was the Compendium of Pharmaceuticals and Specialties (CPS), electronic version, the Micromedex™ Drug Information System or the American Hospital Formulary Service™ (AHFS) Drug Information reference. A summary is provided below for the one ADR not described in the Canadian product monograph.

Reported	Duplicate	Excluded	Pending	Confirmed
40	2	2	8	28

	2014 (n=28)	2013 (n=21)	2012 (n=31)	2011 (n=31)	2010 (n=34)	2009 (n=51)	2008 (n=40)
Up to 5 years	8	10	3	10	6	14	15
6 to 12 years	13	6	14	7	16	16	14
13 to 17 years	7	5	14	14	11	21	11
Not reported	0	0	0	0	1	0	0

An infant passed away several weeks after starting propranolol for hemangioma treatment. Prior to starting propranolol, the child's heart rate and blood pressure were in the normal range and endoscopy of the airway

## Surveillance Studies in 2014

had been performed with no abnormalities noted. Potentially fatal reactions are not listed in available propranolol product monographs (reference: Health Canada's Drug Product Database online, accessed 2015-03-18) with the exception of the monograph for Inderal®-LA (date of revision 2012-09-14) which notes a risk of fatal bronchospasm in patients with asthma. As of 2014-12-31, Health Canada's Canada Vigilance Program has received 19 cases of adverse reactions suspected with the use of propranolol in patients 18 years of age or less.

The other fatal case was the sudden, unexpected death of a young teen on risperidone (Risperdal®), guanfacine (Intuniv XR™), and melatonin. Risperidone was prescribed for explosive temper, aggression, and tics. Guanfacine, an alpha 2a adrenergic receptor agonist, was started several months prior for impulsivity and inattention not controlled with stimulants. The adolescent was concurrently taking melatonin as needed for sleep. He had been admitted to hospital in the recent past for suspected seizures. A few days preceding his death, he had seizure-like episodes. Cardiac arrhythmia was suspected as the cause of death and potentially the cause of the seizure-like episodes. Sudden death and cardiac arrhythmias, including ventricular tachycardia, atrioventricular block, QT prolongation, and torsades de pointes are listed as rare adverse reactions in the Canadian product monograph for Risperdal®/ Risperdal M-Tab® (date of revision 2014-11-06). The Intuniv XR™ Canadian product monograph (date of revision 2015-02-24) describes a risk of hypotension, bradycardia, and syncope under Cardiovascular Warnings and Precautions and includes a caution if using guanfacine in combination with other drugs known to have effects on heart rate or QT prolongation.

Reason for seriousness	2014 (n=28)	2013 (n=21)	2012 (n=31)	2011 (n=31)	2010 (n=32)	2009 (n=45)	2008 (n=35)
Death	2	0	1	1	0	1	3
Life-threatening	9	6	11	11	6	14	12
Hospitalization	19	19	20	18	19	28	18
Disability	3	1	2	4	2	3	0
Medically important condition*	10	7	13	17	18	21	12

\* A medically important reaction is defined as one that may not be immediately life-threatening or result in death or hospitalization but may jeopardize the patient or may require intervention to prevent one of these other outcomes from occurring.

### Suspect health products

Table 4 lists the 26 health products suspected in the 28 cases, sorted by the number of reports received per individual product. The classes of health products most frequently suspected of causing ADRs were:

- 6 cases each – systemic antibacterials, antiepileptics, and cardiovascular drugs (includes 3 reports with guanfacine, an alpha 2a adrenergic receptor agonist, which has an indication for the treatment of attention deficit hyperactivity disorder);
- 3 cases each – psycholeptics, psychoanaleptics, and systemic corticosteroids;
- 2 cases each – immunosuppressants and analgesics;
- 1 case each – antineoplastics, alimentary drugs, and thyroid medications.

Suspect health product	# of times reported
Amoxicillin, guanfacine, phenytoin	3 each
Methylphenidate, morphine, phenobarbital	2 each
Atomoxetine, clarithromycin, clobazam, clonidine, co-trimoxazole*, dexamethasone, infliximab, levofloxacin, lisdexamfetamine dimesylate, lorazepam, mesalazine, methimazole, methotrexate, potassium chloride, propranolol, risperidone, sodium phosphates (enema), sulfasalazine, tacrolimus, valproic acid	1 each

\* Combination product containing two active ingredients

In 23 cases, a single product was suspected of causing the ADR, in 4 cases two suspected products were involved, and in 1 case three suspected products were involved.

Medications used to treat illness in children are increasing and the safety and efficacy of these medications may be different for children than adults. As such, Health Canada recognizes the need to strengthen information related to medications used by children. The ongoing sharing of drug safety information through reporting

CPSP surveillance of ADRs is valuable to Health Canada as it provides information on the benefit-risk profile of medications used by children and has resulted in risk mitigation measures when the risks are deemed to outweigh the benefits.

**Caveat:** *Adverse reactions (ARs) to health products are considered to be suspicions, as a definite causal association often cannot be determined. Spontaneous reports of ARs cannot be used to estimate the incidence of ARs because ARs remain under-reported and patient exposure is unknown.*

### Conclusion

- Systemic antibacterials, antiepileptics, and cardiovascular drugs were the most frequently reported drug classes suspected of causing ADRs in 2014.
- Systemic antibacterials have been the most frequently reported drug class since CPSP surveillance for ADRs began in 2004 with antiepileptic and psychoanaleptic drugs being the second and third most frequently reported. Amoxicillin, carbamazepine, and methylphenidate have been the most frequently reported drugs in each of these three classes.
- The second fatal case scenario describes the hazards of using multiple drugs that prolong QT and the necessity of doing ECG prior to initiation of QT prolonging medication.

### References

Available upon request from the CPSP office

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## Childhood Lyme disease

July 2014 to June 2017

JM Langley, NH Ogden, M Barton, J Konan Koffi, EK Leonard, LR Lindsay

### Highlights 2014

- In the first six months of the study, 24 reports of Lyme disease in children were submitted, of which 17 were confirmed or probable cases.
- The median age of confirmed or probable cases was 6.5 years, with a range of 1 to 16 years.
- Of the 17 confirmed or probable cases, 10 were diagnosed in Nova Scotia and the rest were in Manitoba, Ontario, and Quebec.
- Sixteen of the 17 cases resided in or visited a Lyme endemic area within 30 days of developing symptoms.

### Background and objectives

The complete protocol can be accessed at [www.cpsp.cps.ca/surveillance](http://www.cpsp.cps.ca/surveillance).

### Case definition

A patient less than 16 years of age with Lyme disease, meeting the following criteria:

**Confirmed Lyme disease** – Patient fulfills one of two conditions:

1. Clinical evidence of illness with laboratory confirmation
  - a. Isolation of *Borrelia burgdorferi* from an appropriate clinical specimen
  - OR
  - b. Detection of *B burgdorferi* DNA by PCR in appropriate tissues
2. Clinical evidence of illness with a history of residence in, or visit to, an endemic area\* and with laboratory evidence of infection
  - Positive serologic test using the two-tiered serological approach (i.e., ELISA followed by Western blot assays)

**Probable Lyme disease** – Patient fulfills one of two conditions:

1. Clinical evidence of illness without a history of residence in, or visit to, an endemic area\* and with laboratory evidence of infection
  - Positive serologic test using the two-tiered serological approach (i.e., ELISA followed by Western blot assays)
2. Clinician-observed erythema migrans without laboratory evidence but with history of residence in, or visit to, an endemic area\*

### Exclusion criteria

Confirmation of infection with a non-tick-borne disease, which fully explains symptoms

Cases diagnosed by methods and/or laboratories not recommended by the Public Health Agency of Canada or the US Centers for Disease Control and Prevention

\* An endemic area is defined as a locality in which reproducing populations of *Ixodes scapularis* or *Ixodes pacificus* tick vectors are present and transmission of *B burgdorferi* occurs at the location.

### Results

Lyme disease is a multisystem illness caused by *B burgdorferi*, a spirochete transmitted by ticks (*Ixodes scapularis* in Eastern and Midwestern United States and Central and Eastern Canada, and *Ixodes pacificus* in the Western United States and in British Columbia, Canada). Infection in children can present in the weeks following a tick bite with a characteristic rash called erythema migrans, or later, as heart, joint, skin, or nervous system illness. While Lyme disease is the most commonly reported vector-borne disease in North America, accurate estimates of the burden of illness in Canada

Reported	Duplicates	Excluded	Pending	Confirmed/ Probable
24	2	1	4	17



are not available. Lyme disease is predicted to become more common as the vector tick populations spread further into parts of southern British Columbia, Manitoba, southern Ontario, Quebec, New Brunswick, and Nova Scotia.

Understanding the epidemiology and clinical presentation of childhood Lyme disease in Canada is important because children, particularly boys and those aged 5 to 15 years, represent a risk group for Lyme disease.

The CPSP childhood Lyme disease study was launched in July 2014, and six months of surveillance data are available. Of the 17 confirmed or probable cases, 10 (59%) were from Nova Scotia (one being a US citizen), and 7 (41%) cases were reported from Manitoba, Ontario, and Quebec.

The median age of the 17 confirmed or probable cases was 6.5 years, with a range of 1 to 16 years of age at the time of diagnosis. Eight (47%) cases were male. The age pattern and sex ratio are consistent with that observed in the United States. Eleven (65%) cases were diagnosed in July and August, and 15 (88%) cases resided in or visited a Lyme endemic area in Canada within 30 days of developing symptoms. Of the remaining two cases, one visited or resided in Lyme endemic areas in the United States, and one case may have visited a Lyme endemic area in the United States. Eleven (65%) cases had single or multiple erythema migrans lesions, and fever was the reported symptom in 10 (59%) cases.

### Conclusion

- In 2014, 17 confirmed or probable cases of Lyme disease were reported from four provinces (Manitoba, Ontario, Quebec, and Nova Scotia).
- The median age of confirmed or probable cases was 6.5 years and eight (47%) of the reported cases were male, which is consistent with previous estimates from the United States and Canada.
- Nearly all of the cases resided in or visited a Lyme endemic area in the 30 days prior to developing symptoms.
- The Lyme disease study will continue until 2017 and will help define the presentation, management, and outcomes of this emerging infection affecting Canadian children.

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## Childhood tuberculosis

October 2013 to September 2016

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### Highlights 2014

- Fifty-one confirmed cases of childhood tuberculosis (TB) were identified in 2014; 28 were Aboriginal children and 10 were born overseas.
- Existing data suggest that childhood TB is not distributed evenly across the country. There are some areas and populations with very high rates while others are very low.
- The incidence of both multi-drug-resistant (MDR) TB and extensively drug-resistant TB is increasing in many parts of the world and is present in Canada. One case of MDR-TB was reported in a Canadian child in 2014.

### Background and objectives

The complete protocol can be accessed at [www.cpsp.cps.ca/surveillance](http://www.cpsp.cps.ca/surveillance).

### Case definition

A new active or re-treatment case of TB disease in a patient under the age of 15 years

#### Proven TB disease

##### 1. Laboratory-confirmed

Isolation of *Mycobacterium tuberculosis* complex from any clinical specimen:

Positive culture **or** positive nucleic acid amplification test (NAAT), specifically *M tuberculosis*, *M africanum*, *M canetti*, *M caprae*, *M microti*, *M pinnipedii* or *M bovis* (excluding *M bovis* BCG strain)

##### 2. Clinically confirmed

Probable intrathoracic	Probable extrapulmonary – non-pleural
Signs and symptoms, histology suggestive of TB <b>or</b> close contact with an infectious source case	Signs and symptoms, histology or findings on diagnostic radiology consistent with TB
Chest radiography consistent with intrathoracic TB disease	
<b>and at least one of the following:</b> <ul style="list-style-type: none"> <li>• A positive clinical response to anti-TB treatment</li> <li>• Documented exposure to active case of infectious <i>M tuberculosis</i></li> <li>• Immunological evidence of <i>M tuberculosis</i> infection: Positive TB skin test (TST) <b>or</b> positive interferon gamma release assay (IGRA)</li> </ul>	

#### Presumed TB disease

Treatment for suspected TB disease at any site with at least three anti-TB drugs

#### Cases are identified as “new” or “re-treatment” based on the following criteria:

- **New active case of tuberculosis disease:** No documented evidence or history of previously active tuberculosis
- **Re-treatment case of tuberculosis:**
  1. a) Documented evidence or adequate history of previously active TB that was declared cured or treatment completed by current standards and
    - b) At least a six-month interval since the last day of previous treatment and
    - c) Diagnosis of a subsequent episode of TB that meets the active TB case definition
  - or**
  2. a) Documented evidence or adequate history of previously active TB that cannot be declared cured or treatment completed by current standards and
    - b) Inactive disease for six months or longer after the last day of previous treatment and
    - c) Diagnosis of a subsequent episode of TB that meets the active TB case definition

## Exclusion criteria

- Isolation of another pathogen, including atypical mycobacteria
- Patient arriving in Canada on TB treatment for presumed TB but for whom treatment is stopped because subsequent work-up in Canada suggests no TB
- Patient with latent TB (TST- or IGRA-positive but no clinical or radiologic abnormality)

## Results

TB is caused by the bacterium *Mycobacterium tuberculosis* and less commonly *Mycobacterium bovis* and remains an infection of tremendous clinical and public health importance in Canada. The diagnosis and management of tuberculosis in children, especially in children under 5 years of age, are often particularly complex. Understanding the epidemiology and clinical history of childhood tuberculosis in Canada is important because of: changes in immigration patterns from tuberculosis endemic regions; an increase in immune compromised children due to underlying disease, treatment or transplants; and rapidly changing patterns of microbial resistance.

Reported	Duplicate	Excluded	Pending	Confirmed
93	5	10	27	51

The first full year of the CPSP childhood tuberculosis study was 2014. Of 93 reported cases, 51 have been confirmed as unique cases. All confirmed cases came from six provinces and territories: Ontario (15), Nunavut (11), Quebec (10), Manitoba (9), and the remaining cases were from British Columbia and Alberta.

The age of confirmed cases was between 2 months and 15 years, with 21 boys and 29 girls (one did not report). Canadian-born children represented 41 (80%) of confirmed cases, while 10 (20%) were born elsewhere. Of Canadian-born children, 18 (44%) were Inuit, 10 (24%) were First Nations, 11 (27%) were non-Aboriginal, and 2 (5%) did not specify.

Overrepresentation of TB in First Nations and Inuit children is notable and is linked to a number of complex social factors that will be further explored upon completion of data collection for this study. These data are preliminary and, as such, there may be differential reporting rates across the country. However, high rates of TB in First Nations and Inuit populations among all age groups have been previously described and, in the most recent PHAC data available (2013), the overall rates of TB were 21.8 and 154.2 cases per 100,000 population respectively.

Forty-nine (96%) cases had intrathoracic involvement with 41 (80%) reporting lung and 6 (12%) reporting pleural involvement. Fourteen (27%) cases reported extrapulmonary involvement, with the most common site being the central nervous system with 10 (20%) cases.

Twenty-seven (53%) cases were microbiologically proven, either by culture or nucleic acid amplification, whereas 21 (41%) were diagnosed clinically. Additionally, two cases were presumptively diagnosed. Of the cases that were culture positive, two showed resistance, with one reporting only isoniazid resistance and the other being classified as MDR-TB, as it was resistant to isoniazid, rifampicin, and rifabutin.

## Conclusion

- Of the 51 confirmed cases of childhood TB in 2014, 28 (55%) were Aboriginal children, 11 were non-Aboriginal Canadian-born children, and 10 were children born overseas.
- Nearly all cases had intrathoracic involvement, while 14 (27%) showed extrapulmonary manifestations.
- Of all culture positive cases, one showed MDR-TB.

## Publications and presentations

Encounters with an old foe: Childhood tuberculosis in Canada. Kitai I, Morris SK. Paediatr Child Health 2014;19(2):89–90

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# Early-onset major depressive disorder

## January 2012 to December 2014 – Final report

D Korczak, M Feldman, J LeBlanc, M Ofner, P Parkin, S Wong

### Highlights

- Twenty-two cases of early-onset major depressive disorder (EOMD) were confirmed with 86% impaired in three or more functional domains, and 82% having one or more psychiatric comorbidities.
- Children with EOMD experienced severe symptoms: 73% reported suicidal thoughts and 23% had attempted suicide.
- Parental history of a mood disorder, most commonly major depressive disorder (MDD), was present in 68% of cases.

### Background and objectives

The complete protocol can be accessed at [www.cpsp.cps.ca/surveillance](http://www.cpsp.cps.ca/surveillance).

### Case definition

A child 5 to 12 years of age seen in the previous month, with newly diagnosed early-onset major depressive episode (EOMD), including a child with unipolar mood disturbances sufficient to cause a disruption to social, family, and/or academic functioning

“Major depressive episode” is defined in DSM-IV-TR as:

- 1) Depressed or irritable mood, most of the day, nearly every day,

**or**

- 2) Markedly diminished interest or pleasure in all, or almost all, activities most of the day,

which is either newly present or has clearly worsened compared with the child’s pre-episode status,

**and**

At least **four** of the following seven symptoms present during the same two-week period as either (1) **or** (2) above. These symptoms occur daily or near daily and represent a distinct change from previous functioning.

- 1) Significant weight change, failure to make expected weight gains **or** significant appetite change
- 2) Insomnia (difficulty falling asleep, night-waking or waking too early) **or** hypersomnia
- 3) Psychomotor agitation **or** retardation: observable by others and does not represent subjective feelings
- 4) Fatigue **or** loss of energy
- 5) Feelings of worthlessness **or** excessive or inappropriate guilt (not merely guilt about being sick)
- 6) Diminished ability to think or concentrate, **or** indecisiveness
- 7) Recurrent thoughts of death, recurrent suicidal ideation, **or** a suicide attempt

**and**

Impairment in social functioning (social withdrawal, family or peer conflicts) or academic functioning (school refusal, decreased school performance), which is either newly present or worsened compared with pre-episode status

### Exclusion criteria

- 1) Symptoms due to the direct physiological effects of a substance or a general medical condition
- 2) Symptoms occurring exclusively during acute bereavement period (within two months after the loss of a loved one). Note: This exclusion does not apply to palliative care patients
- 3) A previous diagnosis of a manic episode or bipolar disorder

### Results

After three years of surveillance, 45 EOMD cases were reported and 11 were excluded: 10 due to age older than 12 years, and 1 due to revised diagnosis by a child psychiatrist. Of the 22 confirmed cases, 11 (50%) were

boys. Most children (16, 73%) with childhood-onset depression were globally impaired in all functional domains.

Eighteen children (82%) with EOMD reported distressing symptoms for more than 6 months and 11 of these for more than 12 months prior to presentation. Fifteen (68%) cases had a parental history of mood disorder, most

commonly maternal MDD (11, 50%), consistent with existing literature. Psychiatric comorbidity was present in 18 (82%) cases, most commonly attention deficit hyperactivity disorder (ADHD) and anxiety, indicating very early onset of difficulties with affective and behavioural regulation among these children. Children experienced severe depressive symptoms: 16 (73%) children reported suicidal ideation and 5 (23%) children had attempted suicide. Most cases were receiving treatment: 17 (77%) with medication, 14 (64%) with non-pharmacological treatment, and 11 (50%) with combination medication and psychotherapy. Fourteen of the 17 (83%) cases receiving pharmacological treatment for depression were treated with a selective serotonin reuptake inhibitor (SSRI): 7 cases were treated with fluoxetine, 3 were treated with escitalopram, 2 were treated with citalopram, and 1 each with sertraline and paroxetine. Three children were treated with amitriptyline, two were treated with lamotrigine, and one child each was treated with venlafaxine or valproic acid. Two children were treated with quetiapine, an atypical antipsychotic medication. Eleven (50%) patients were treated with two or more medications.

Results from this study are consistent with the literature in suggesting that early-onset depression is a familial, debilitating illness that may be present for a prolonged period of time prior to presentation. Increased knowledge regarding the incidence and presentation of children affected with EOMD is important in designing effective diagnostic and management approaches for children with this treatable illness.

## Conclusion

- Most children with EOMD were globally impaired in all functional domains.
- Psychiatric comorbidity was present in nearly all cases, most commonly ADHD and anxiety.
- Children with EOMD are frequently treated with anti-depressant medication, either singly or in combination with psychotherapy, and polypharmacy is common.

## Publications and presentations

Korczak DJ. Identifying depression in childhood: Symptoms, signs and significance. *Paediatr Child Health* 2012;17(10):572

### References

Available upon request from the CPSP office

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Year	Reported	Duplicate	Excluded	Pending	Confirmed
2012	20	1	6	2	11
2013	9	0	3	3	3
2014	16	0	2	6	8
<b>Total</b>	<b>45</b>	<b>1</b>	<b>11</b>	<b>11</b>	<b>22</b>

## Fragile X syndrome

### April 2012 to March 2014 – Final report

G Aubertin, J Down, G Graham, T Nelson, M Ofner, C Paribello

#### Highlights

- Thirty cases of fragile X syndrome (FXS) were confirmed during the study period.
- The average age at postnatal diagnosis was 4 years and 2 months.
- A family history of FXS or an FX-related condition was present in a minority of cases (13%), of which four cases had a delayed diagnosis (ages 5, 5, 6, and 9 years).
- Co-morbid diagnoses included speech and communication problems, and autism.

#### Background and objectives

The complete protocol can be accessed at [www.cpsp.cps.ca/surveillance](http://www.cpsp.cps.ca/surveillance).

#### Case definition

A new patient less than 18 years of age with diagnosed fragile X syndrome meeting the following criteria:

- 1) **Genetic criteria:** Males or females, with laboratory confirmation of a CGG-repeat allele in the full mutation size range (>200 repeats), including mosaicism

and

- 2) **Clinical criteria, one** of the following:
  - Global developmental delay, manifesting as the clinical impression of delays in two or more domains of development **or**
  - Intellectual disability, mild, moderate or severe, diagnosed through standardized psychological testing **or**
  - Asymptomatic infant, tested because of a positive family history, including prenatally diagnosed cases

#### Exclusion criteria

Clinical evidence of global developmental delays or intellectual disability with laboratory confirmation of a CGG-repeat allele in the normal or premutation size range

#### Results

Over the duration of the study, there were 41 reported cases, of which 30 have been confirmed, and the detailed questionnaire completion rate at the time of analysis was 81%. Twenty-six (87%) cases were male, 3 (10%) were female, and one (3%) was not specified. The average age at diagnosis was 4 years and 2 months (range: 15 months to 9 years 11 months), which is in keeping with published data. In contrast, the average age at parental first concern was 21 months; and 16 (53%) cases were reported with first concerns at or below 18 months (range: 2 months to 60 months).

Year	Reported	Duplicate	Excluded	Pending	Confirmed
2012*	14	1	0	3	10
2013	21	0	3	4	14
2014†	6	0	0	0	6
<b>Total</b>	<b>41</b>	<b>1</b>	<b>3</b>	<b>7</b>	<b>30</b>

\* April 1 to December 31, 2012

† January 1 to March 31, 2014

Where location information was provided, 12/25 (48%) of the confirmed cases were from Ontario, 5/25 (20%) cases were from Manitoba, and 8/25 (32%) cases were from the remaining Western Canada provinces (BC, AB, and SK). There were no cases reported from Atlantic Canada.

The ethnicity of the majority of cases was Caucasian (22, 73%), but diversity was seen with a combination of differing ethnicities in a minority of cases, including Asian, mixed East Indian/Middle Eastern/Caucasian, First Nations, and Black.

Four (13%) of the cases had a known family history of FXS or an FX-related condition. All four of these cases had a delayed diagnosis (two at age 5, one at age 6, and one at age 9 years 11 months). In the remaining 87% of cases, there was either no family history of FXS or FX-related conditions, or this was reported as unknown or left blank.

Developmental delays were the presenting concern in 25 (83%) cases, while in 8 (27%) cases autism was the reason for initial assessment. Speech and communication problems were reported in 23 (77%) cases. Other comorbidities included recurrent otitis media in 8 (27%), ADHD in 7 (23%), sleep apnea in 3 (10%) cases, and seizures in only one (3%) case.

### Discussion

There is limited information in the medical literature on the demographics of FXS in Canada. This study aimed to ascertain the minimum incidence of new FXS diagnoses, along with informing demographics, clinical features, burden of illness, and management approaches in Canada. Results of this study indicate that the age at diagnosis, ethnicity, geographic distribution, and medical comorbidities are generally within the ranges expected for the newly diagnosed FXS population. Notable is that no cases were reported from Atlantic Canada, and the disproportionately high number of cases in Manitoba (20% of total cases, compared to Manitoba's 3% of the Canadian population). A low prevalence of FXS in the Maritimes has previously been described. The frequency and range of associated medical concerns is consistent with what has been previously described in the literature but may be under-reported for two reasons: first, many of the children were reported to the CPSP with a new diagnosis of FXS before they had been fully evaluated for possible complications; and second, the results of these assessments were not always available to reporting physicians.

This study identified evidence of delayed diagnoses based on the early age of parents' reported first concern and the age at diagnosis. Delayed diagnoses have been a consistent problem in the United States and this concern has led to recommendations for population-based screening from some of the experts in the field of fragile X-related conditions. This study has shown that the experience of Canadian families affected by fragile X-related disorders is in many ways similar to that of American families, and that an improved approach to recognizing and diagnosing FXS would be of benefit. A good understanding of the characteristics of the FXS patient population in Canada is essential, given two recent developments in the field: enhanced newborn screening technology and novel pharmaceutical agents in clinical trials, giving rise to speculations that targeted therapies will be available to patients in the next few years.

### Conclusion

- Geographic and ethnic distributions of diagnoses of FXS in Canada tend to reflect the Canadian population and are in keeping with international prevalence estimates.
- In 87% of cases, there was no known family history of FXS or FX-related conditions, or family history was unknown or left blank.
- Delayed diagnoses were common, even with a family history of FXS or FX-related condition.

### Publications and presentations

Aubertin G. Fragile X syndrome: Are paediatric health care providers missing the diagnosis? *Paediatr Child Health* 2013;18(5):251-2

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## Hypoglycemia in low-risk term newborns

April 2014 to March 2016

M Flavin, K Grewal, K Coughlan, A Gallipoli, K Gregoire, L Hu, JA León, H Osiovich, J Ray, M Sgro

### Highlights 2014

- Of the 25 confirmed cases of hypoglycemia in low-risk term newborns reported in the first nine months of surveillance, 28% were born by emergency Caesarean section and 38% needed some resuscitation at birth.
- The majority (56%) of cases presented in the first six hours of life. Those who had hypoglycemia recognized beyond six hours after birth had more morbidity.
- A brain MRI was done in 29% of cases, many having MRI evidence of hypoglycemic brain injury.
- Five cases had a seizure at the time of presentation. Only one case had abnormal neurologic signs at the time of hospital discharge.

### Background and objectives

The complete protocol can be accessed at [www.cpsp.cps.ca/surveillance](http://www.cpsp.cps.ca/surveillance).

### Case definition

Any otherwise healthy neonate less than 96 hours (4 days) old with all of the following:

- Term gestation: 37–42 weeks
- Birth weight: 2500–3999 grams
- Hypoglycemia, defined as whole blood or serum glucose <2.0 mmol/L
- Hypoglycemia treated with IV dextrose

### Exclusion criteria

Neonate being monitored for hypoglycemia because of known risk factors, i.e., maternal diabetes (gestational or pre-gestational), growth restriction, macrosomia, or important neonatal illness

### Results

In the first nine months of the study, 64 cases of hypoglycemia in low-risk term newborns were reported and 25 cases were confirmed. Five cases did not meet the case definition and 34 detailed questionnaires were pending at the time of analysis.

Reported	Duplicate	Excluded	Pending	Confirmed
64	0	5	34	25

Where indicated on the detailed questionnaire, sex in the 25 cases was equally distributed. The mean birth weight was 3277 g (SD ± 416) and the mean gestational age at birth was 39 weeks (SD ± 1.4). Where ethnicity information was provided, 3/17 (18%) cases were from First Nations families.

Fourteen (56%) cases of hypoglycemia presented in the first six hours of life. Seven of the 25 (28%) cases were born by emergency Caesarean section and meconium was present at delivery in 7/24 (29%) cases. Resuscitation was required at birth in 9/24 (38%) cases, and in 7/9 (78%) of those cases, the duration of resuscitation was under two minutes. Feeding issues were reported in 6/21 (29%) cases and 3/17 (18%) cases had temperature under 36.5°C before they received IV dextrose. A prior glucose measurement was done in 6/21 (29%) cases; 2 cases were monitored as part of a local monitoring protocol which included newborns requiring positive pressure ventilation at birth, and the other 4 cases had clinical signs suggestive of hypoglycemia. For the remaining 15/21 (71%) cases which did not have prior glucose measurement, a variety of factors triggered the first measurement including 7 cases with jitteriness, 5 with cyanosis/desaturation, and 5 with suspected or definite seizure. Initiation of IV dextrose was prompted by concerning glucose levels in 23/25 (92%) cases, concerning signs in 14/25 (56%), and unsuccessful attempts at normalization of glucose levels in 7/25 (28%).

A workup for an underlying cause for the hypoglycemia was done in 14/25 (56%) cases, of which 9/14 (64%) had an identified underlying diagnosis or confounding factor. Brain imaging was done in 10/24 (42%) cases. Seven



of 24 (29%) cases had a brain MRI, all of which were abnormal: 5/7 (71%) had MRI findings consistent with or suggestive of hypoglycemic brain injury, of which 4/5 (80%) had hypoglycemia recognized more than six hours after birth.

In terms of short-term outcomes, 7/24 (29%) cases were given an uncertain prognosis at hospital discharge and only one case had abnormal neurological signs at the time of discharge.

### Conclusion

- Twenty-five cases of hypoglycemia in low-risk term newborns were confirmed during the first nine months of the study.
- Approximately one third of cases had an emergency Caesarean section, meconium, or brief neonatal resuscitation suggestive of perinatal distress.
- Many low-risk newborns with hypoglycemia had evidence of brain injury particularly those with hypoglycemia recognized more than six hours after birth.
- Additional case ascertainment over this two-year surveillance study will be important to confirm these findings and identify other factors associated with hypoglycemia in low-risk newborns.

### Publications and presentations

Flavin M, Grewal K, Hu L. Hypoglycemia in newborns with no pre-identified risk factors. Canadian Paediatric Society Annual Conference, Montreal, June 2014. (Poster presentation)

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## Kawasaki disease

### November 2013 to November 2014 – Final report

R Scuccimarri, R Yeung, D Cabral, N Dahdah, DG Human, HA Hume, B Lang, B McCrindle, S Schwartz

#### Highlights

- The study confirmed 285 cases of Kawasaki disease (KD).
- Most patients (93%) received IVIG therapy; of these, 25% failed initial therapy and 11% developed evidence of IVIG-associated hemolysis.
- Most patients recovered without cardiac sequelae with 5% developing coronary artery aneurysm and 23% developing dilatation sometime during their disease course.
- Study results may have important implications for disease management and patient advocacy.

#### Background and objectives

The complete protocol can be accessed at [www.cpsp.cps.ca/surveillance](http://www.cpsp.cps.ca/surveillance).

#### Case definition

A new patient presenting before the age of 18 years with a definite or presumed diagnosis of Kawasaki disease:

- 1) **Complete KD**, defined as fever persisting for five\* days or more

AND the presence of **at least four** of the following clinical criteria:

- Changes in the peripheral extremities
  - erythema of the palms and/or soles; edema of the hands and/or feet; periungual desquamation
- Polymorphous rash
- Bilateral bulbar conjunctival injection without exudate
- Changes in the lips and oral cavity
  - erythema and/or cracking of the lips; strawberry tongue; diffuse erythema of the oropharynx
- Cervical lymphadenopathy > 1.5 cm diameter, usually unilateral

\* Presumptive diagnosis and initiation of treatment may be made before the fifth day of fever.

- 2) **Incomplete KD**, defined as fever of five days or more and less than four clinical criteria

- 3) **Other KD**, defined as KD not fulfilling criteria for complete or incomplete KD but presumed because of a feature on echocardiogram or follow-up (i.e., periungual desquamation) that has led the treating physician to recommend treatment and/or cardiac follow-up

#### Results

During the period of surveillance, 453 cases of acute Kawasaki disease were reported, and 285 of these were confirmed as individual cases (Table 1). Of the 285 cases, 174 (61%) were male. The median age at diagnosis was 3.4 years (range: 0.1 to 17.9 years). One hundred and seventy-seven (62%) cases had complete KD and 91 (32%) had an incomplete presentation. Paediatric rheumatologist members of CAPRI (Canadian Alliance of Paediatric Rheumatology Investigators) also reported cases to the CPSP.

**TABLE 1 – KD cases from November 1, 2013 to November 30, 2014**

Reported	Duplicate	Excluded	Pending	Confirmed
453	100	15	53	285

Ethnicities were reported as follows (percentages exceed 100% due to some cases having more than one ethnic origin): 127 (45%) cases were Caucasian, 62 (22%) were Asian, 21 (7%) were Black, 17 (6%) were South Asian, 13 (5%) were Middle Eastern, 5 (2%) were Latin American, 6 (2%) were Aboriginal, 7 (2%) were reported as 'other,' and in 44 (15%) cases, ethnicity was not documented. One hundred and forty-six (51%) cases were from Ontario, 55 (19%) cases were from Quebec, 34 (12%) were from British Columbia, and the remaining 50 (18%) were from the rest of Canada. Most patients (264, 93%) received IVIG therapy during the acute phase of disease.

Of these, 66 (25%) received more than one dose. Of the 264 children who received IVIG therapy, 67 (25%) experienced an adverse effect including hemolysis in 29 (11%) cases (Table 2).

Thirteen of the 285 (5%) cases developed a coronary artery aneurysm and 65 (23%) developed a dilatation sometime during the course of disease.

Kawasaki disease and its treatment complications are a significant public health concern. The results of this study provide much needed evidence to allow for the development of management guidelines to improve treatment and to allow for advocacy for improved therapies.

TABLE 2 – IVIG-related side effects		
	Number of cases	Percentage of cases n=264
Cases experiencing a side effect	67	25%
<b>Individual side effects</b> (one or more for each case)		
Allergic reaction	16	6%
Mild-moderate headache	14	5%
Suspected aseptic meningitis	7	3%
Renal impairment	0	0%
Thrombosis	0	0%
Hemolysis	29	11%
Other	12	5%

## Conclusion

- KD was reported from across Canada and spanned ethnicities.
- IVIG was used in most of the confirmed cases of KD.
- The rate of IVIG non-response and IVIG-associated complications including hemolysis were higher than previously reported in the literature.
- Children developed coronary artery aneurysms at a rate similar to that found in previous North American studies.

## Publications and presentations

Scuccimarri R, Yeung RSM. Kawasaki disease: High index of suspicion needed in a febrile child. *Paediatr Child Health* 2014;19(5):239-40

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## Periodic fever syndromes

### September 2011 to August 2014 – Final report

P Dancey, S Benseler, M Gattorno, AK Junker, RM Laxer, P Miettunen, LA Turner

#### Highlights

- Over this three-year study, 179 cases of periodic fever syndromes (PFS) were confirmed.
- Periodic fever, aphthous stomatitis, pharyngitis, and adenitis (PFAPA) was the most frequently reported diagnosis, followed by undefined PFS, and familial Mediterranean fever (FMF).
- On average, symptom onset occurred 2.5 years before diagnosis and patients saw multiple physicians before diagnosis.

#### Background and objectives

The complete protocol can be accessed at [www.cpsp.cps.ca/surveillance](http://www.cpsp.cps.ca/surveillance).

#### Case definition

A patient less than 18 years of age presenting with a newly diagnosed periodic fever syndrome (autoinflammatory syndrome) meeting the criteria outlined below

#### Inclusion criteria

The patient must have one of the following diagnoses (see appendix and table in protocol for specific details and characteristic features):

- Familial Mediterranean fever (FMF)
- Tumor necrosis factor receptor-associated periodic syndrome (TRAPS)
- Hyperimmunoglobulinemia D syndrome (HIDS)
- Cryopyrin-associated periodic syndromes (CAPS), including familial cold autoinflammatory syndrome (FCAS), Muckle-Wells syndrome (MWS), and neonatal-onset multisystem inflammatory disease (NOMID)
- Periodic fever, aphthous stomatitis, pharyngitis and adenitis (PFAPA)
- Periodic fever syndrome – undefined

#### Exclusion criteria

- Detailed clinical assessment and investigations compatible with infections, malignancy, or the classical inflammatory or autoimmune rheumatic diseases (e.g., systemic lupus erythematosus, systemic juvenile idiopathic arthritis, inflammatory bowel disease)
- Febrile attacks with regular periodicity and low neutrophil counts, suggestive of cyclic neutropenia

#### Results

During the three-year surveillance period, 221 cases of PFS were reported. The overall detailed questionnaire completion rate was 87%. Of the 179 confirmed cases, 84 (47%) were PFAPA, 72 (40%) were undefined PFS, 17 (10%) were FMF, and the remaining 6 cases were CAPS, TRAPS, and HIDS (Table 2). The mean age at diagnosis was 5.5 years (range: 12 months to 16 years). On average, symptom onset occurred 2.5 years before diagnosis (range: 1 to 4 years). The delay was longest for the few patients with CAPS and TRAPS. The majority of reporting physicians were rheumatologists (58%) and paediatricians (36%). The remaining 6% included specialists from infectious disease, nephrology, gastroenterology, emergency medicine, hematology, and endocrinology. Patients had seen multiple physicians prior to their diagnosis (average 2.7, range: 1 to 9). Cases were identified in all provinces across Canada with the majority (103, 58%) from Ontario. Clinical characteristics of cases are described in Table 2.

Year	Reported	Duplicate	Excluded	Pending	Confirmed
2011*	27	1	4	0	22
2012	81	0	6	16	59
2013	67	2	0	7	58
2014†	46	0	0	6	40
<b>Total</b>	<b>221</b>	<b>3</b>	<b>10</b>	<b>29</b>	<b>179</b>

\* September 1 to December 31, 2011

† January 1 to August 31, 2014

TABLE 2 – Clinical characteristics of 173/179 confirmed cases

PFS type	Number of cases	Sex	Mean age at diagnosis (range)	Mean age at symptom onset (range)	Number of fever attacks per year (range)	Mean duration of attacks (range)	Mean duration of fever-free intervals (range)
PFAPA	84	43 females 38 males 3 not stated	4 years (12 months–11 years)	3 years (6 months–10 years)	10 (4–18)	4 days (1–8)	32 days (17.5–105)
Undefined	72	24 females 43 males 5 not stated	5 years (19 months–16 years)	3 years (6 months–15 years)	12 (3–30)	4 days (1–10.5)	30 days (3–135)
FMF	17	9 females 8 males	5 years (2–11 years)	3 years (6 months–8 years)	13 (6–24)	3 days (1–8)	24 days (10–49)
CAPS, TRAPS and HIDS*	6						

\* In accordance with CPSP policy, detailed clinical and genetic characteristics are not reported when low case numbers are identified.

For the 84 cases of PFAPA, the most frequent fever-associated manifestations were pharyngitis (75, 89%), stomatitis (51, 61%), cervical lymphadenopathy (50, 60%), fatigue (38, 45%), abdominal pain (26, 31%), vomiting (21, 25%), headache (20, 24%), myalgias (18, 21%), and arthralgias (17, 20%).

For the 72 cases of undefined PFS, the most frequent fever-associated manifestations were fatigue (34, 47%), abdominal pain (26, 36%), arthralgias (26, 36%), pharyngitis (25, 35%), lymphadenopathy (19, 26%), vomiting (17, 24%), headache (16, 22%), stomatitis (14, 19%), maculopapular rash (11, 15%), myalgias (10, 14%), and diarrhea (9, 13%). Of the undefined PFS cases, 36% were felt to be PFAPA-like, 10% FMF-like, 5% HIDS-like, 4% CAPS-like, and 1% TRAPS-like. These cases with the “like” designation had features of the condition but either did not meet the full criteria, or confirmatory tests were pending at the time of the reporting. The remaining undefined PFS cases met the criteria for a PFS but were not felt to be similar to other known subtypes.

For the 17 cases of FMF, the most frequent fever-associated manifestations were abdominal pain (12, 71%), arthralgias (8, 47%), pharyngitis (6, 35%), fatigue (6, 35%), diarrhea (5, 29%), headache (4, 24%), myalgias (4, 24%), vomiting (3, 18%), and stomatitis (2, 12%). Ten of the patients had genetic mutations identified in the *MEFV* gene, the majority being M694V, in addition to V726A, M680I, E148Q, and M694I. Seven patients who had at least a single mutation in the *MEFV* gene, or in whom genetic testing was not done or pending, met the Tel Hashomer clinical criteria for the FMF diagnosis.

Of the CAPS cases, two were diagnosed as having MWS. The most common disease manifestations for CAPS included conjunctivitis, urticarial rash, and arthralgias. One case also reported headache, abdominal pain, pharyngitis, and enthesitis. Of the TRAPS cases, patients reported arthralgias and abdominal pain. Additional fever-associated manifestations included myalgias, headache, oligoarthritis, diarrhea, and maculopapular rash. For HIDS, fever-associated manifestations included a maculopapular rash, pharyngitis, abdominal pain, vomiting, diarrhea, myalgias, and conjunctivitis.

Among all cases reported in the study, 58% had genetic testing completed as part of their diagnosis. The majority of cases without any genetic testing had been diagnosed with PFAPA, which is diagnosed on clinical grounds unless genetic testing is needed to exclude an alternative PFS.

PFS represent rare forms of autoinflammatory disease, which affect many Canadian children. PFS can be recognised by the fever pattern and other disease manifestations. With the exception of PFAPA, a genetic diagnosis can often be made; however, there were many children who met criteria for a PFS but nevertheless did not fit into the few existing diagnostic categories and were diagnosed as undefined PFS. This CPSP study identified that the most common PFS diagnosed by Canadian paediatricians was PFAPA followed by undefined PFS and FMF. CAPS, HIDS, and TRAPS were rarely diagnosed. Symptom onset tended to occur before age 5 years

and some children presented during the first year of life. Children with PFS were seen by multiple physicians over an average of two to three years before a diagnosis was made. It is hoped that increased awareness of this rare disease will facilitate earlier diagnoses and the initiation of effective treatments for these children.

### Conclusion

- Many Canadian children are affected with PFS each year with frequent attacks of fever.
- Each type of PFS is unique and can be recognized by the periodicity of attacks and fever-associated manifestations.
- The onset of symptoms was often years prior to diagnosis.
- Genetic testing was frequently reported and is important to better define the PFS subtypes, opening the possibility of effective treatments.

### Publications and presentations

Dancey P, Benseler S, Junker AK, Laxer RM, Miettunen P, Turner LA, Gattorno M. The challenge of periodic fevers in children. *Paediatr Child Health* 2012;17(3):123

Dancey P, Benseler S, Gattorno M, Junker AK, Laxer RM, Miettunen P, Turner LA. Periodic fever syndromes in Canada: Results from a national surveillance program. *Pediatric Rheumatology Symposium*, Orlando, in April 2014. (Poster presentation)

### References

Available upon request from the CPSP office

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## Severe alcohol intoxication in adolescents

March 2013 to February 2015

A Acker, K Thomas, D Allain, K Dow, C Korenblum, K Leslie, M Norris, A Vandermorris

### Highlights 2014

- In the second year of this two-year surveillance study, 15 cases of severe alcohol intoxication (SAI) in young Canadian adolescents were identified.
- The mean age was 13.9 years.
- Incidence and presenting features were similar among males and females.
- Average blood alcohol levels were high (mean 2.54 g/L).
- Four adolescents required mechanical respiratory assistance and one died in hospital.

### Background and objectives

The complete protocol can be accessed at [www.cpsp.cps.ca/surveillance](http://www.cpsp.cps.ca/surveillance).

### Case definition

An adolescent who meets the following criteria:

- Between 11 and 15 years of age, inclusively
- Blood alcohol level >0 g/L (if available)
- Presenting with severe intoxication with impaired consciousness requiring prolonged ER observation ( $\geq 6$  hours) or hospital admission

### Results

In the second year of this two-year surveillance study, there were 23 reported cases and 19 completed questionnaires. The detailed questionnaire completion rate at the time of analysis was 83%. Of these, 15 cases met the case definition. There were seven males and eight females with a mean age of 13.9 years. The youngest case was 11 years old.

Reported	Duplicate	Excluded	Pending	Confirmed
23	0	4	4	15

Cases were reported from across the country, although no cases were reported from Alberta, Manitoba, New Brunswick, or the territories. Seven (47%) of the cases were from Ontario and Quebec.

The age of first alcohol use was known and reported in 8/15 (53%) cases; of these, four (50%) were 13 years of age or younger when they first used alcohol, and the youngest age of first use reported was 7 years. A prior emergency room visit related to alcohol use was identified in 3/14 (21%) cases. The mean blood alcohol level of adolescents presenting with severe intoxication was 2.54 g/L (range: 1.52 to 4.70). There did not appear to be a difference in blood alcohol levels between sexes.

The most common source of alcohol was friends (9/14, 64%). The majority (12/14, 86%) of adolescents consumed spirits (hard liquor) and 10/14 (71%) consumed alcohol at parties (i.e., outside of the parents' home). Concurrent use of cannabis was common (5/15, 33%).

Just over half (7/13, 54%) of the patients were observed for over 12 hours in the emergency department or as inpatients (two cases not specified). The majority (13/15, 87%) presented between 6 p.m. and 6 a.m. Four adolescents (27%) required mechanical respiratory assistance. One adolescent died from a cardiac arrest after presenting to the hospital with SAI. Follow-up medical care was provided for 11 (73%) cases, the majority with adolescent medicine subspecialty clinics.

## Surveillance Studies in 2014

After almost two years of surveillance, nearly 60 cases of SAI in adolescents have been identified. This is likely only the tip of the iceberg. The young age of presentation (youngest 11 years of age) and severity of illness (25% requiring respiratory assistance and one death) are concerning. Data on Canadian adolescents presenting to the emergency department with SAI will inform strategies for education, prevention, and harm reduction. As the majority of adolescents with severe intoxication consumed spirits, more education for adolescents about the higher potency of spirits relative to other alcoholic beverages and the dangers of consumption, even in small amounts, could reduce the incidence of SAI in children and adolescents. Consideration of more stringent controls on spirits to help decrease adolescents' access would assist with harm reduction.

### Conclusion

- Numerous Canadian adolescents between the ages of 11 and 15 years with SAI presented to emergency departments for observation or were admitted to hospitals across the country with very high blood alcohol levels.
- The young age of presentation and severity of illness are cause for concern.
- Consideration of more stringent controls and education for adolescents about the higher potency of spirits and the risk of SAI may be beneficial.

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## Sudden unexpected death in epilepsy

January 2014 to December 2015

E Donner, M Bhatt, L Carmant, N Jette, I Mohamed

### Highlights 2014

- Five cases of sudden unexpected death in epilepsy (SUDEP) were reported in the first 12 months of surveillance.
- All reported deaths occurred in children with poorly controlled, convulsive seizures.
- The small number of reported cases suggests that Canadian paediatricians are not aware of all SUDEP deaths occurring among children within the community or the frequency of such events is rare.

### Background and objectives

The complete protocol can be accessed through [www.cpsp.cps.ca/surveillance](http://www.cpsp.cps.ca/surveillance).

### Case definition

Sudden unexpected death in a child less than 18 years of age:

- With epilepsy (defined as > 1 unprovoked seizure)
- With or without evidence of a recent seizure
- Without documented status epilepticus
- Without trauma

*Definite SUDEP* is defined as meeting the above criteria, and a post-mortem examination does not reveal a cause of death. *Probable SUDEP* is defined as definite SUDEP, but without autopsy.

### Results

There were five confirmed reports of SUDEP through the CPSP in 2014, and the detailed questionnaire completion rate was 100%. Two deaths met criteria for definite SUDEP, with autopsy failing to determine cause of death; the remaining three deaths were classified as probable SUDEP.

Reported	Duplicate	Excluded	Pending	Confirmed
5	0	0	0	5

Of the five confirmed cases, the age at death ranged from 2 to 16 years with a mean of 8.4 years (SD  $\pm$  5.0). Four (80%) of the children were female. The age of first provoked seizure ranged from 1.5 to 31 months with a mean of 10.7 months (SD  $\pm$  12.6). All children were reported to have had some degree of developmental delay and drug-resistant, poorly controlled, convulsive seizures. One child had a lifetime frequency of 10–100 seizures, three children were reported to have a lifetime frequency of 100–500 primary and secondary generalized tonic-clonic seizures; and another child had a lifetime frequency of more than 500 seizures. Three (60%) children had a history of nocturnal seizures; in the remaining cases, this information was not available.

Four children were reported to be asleep and the death was not witnessed. In the fifth case, the child was reported to be awake at the time of death, with seizures observed prior to death. A coroner or medical examiner was alerted to the death in all cases; however, an autopsy was performed in only three of the five deaths.

Only one family is reported to have been aware of the risk of SUDEP prior to the death of their child.

### Conclusion

- Five cases of SUDEP were reported in the first 12 months of surveillance.
- Based on best available data regarding incidence of SUDEP in children, a minimum of nine cases were expected to occur over 12 months in Canada suggesting that Canadian paediatricians are not aware of all cases of SUDEP among children in the community or that SUDEP is quite rare.

## Surveillance Studies in 2014

- Most deaths occurred in children considered to be at risk of SUDEP with frequent drug-resistant, convulsive seizures, and most occurred during sleep when children were unsupervised.
- Most families of children with SUDEP were not aware of this risk.
- To optimize surveillance of SUDEP in Canadian children, an ongoing parallel study is being conducted in association with the Canadian Pediatric Epilepsy Network, surveying paediatric neurologists across Canada.

### Publications and presentations

Donner EJ. Sudden unexpected death in epilepsy: Who are the children at risk? *Paediatr Child Health* 2014;19(7):389

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#### Principal investigator

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# Survey Questions

## Avoidant/restrictive food intake disorder

February 2014

DK Katzman, ML Norris, K Stevens

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### Highlights

- Avoidant/restrictive food intake disorder (ARFID) is a new diagnostic category in the *Diagnostic and Statistical Manual of Mental Disorders, Fifth Edition (DSM-5)*.
- A one-time CPSP survey revealed that 63% of paediatricians and paediatric subspecialists were unfamiliar with the diagnosis of ARFID.

### Results

ARFID is a new diagnostic category in the section on Feeding and Eating Disorders in the *DSM-5*. It is a new conceptualization of feeding disorder of infancy or early childhood in the *DSM-IV*. The aim of this revision was to: improve the clinical utility by adding more detail to the diagnostic criteria; include individuals with clinically significant eating problems who were assigned a *DSM-IV* diagnosis of eating disorder not otherwise specified; and widen the criteria to be applicable across the lifespan.

Information regarding the frequency of ARFID among children and youth is lacking. The CPSP conducted a national one-time survey to assess the recognition of this new diagnostic category among Canadian paediatricians and to identify the frequency of ARFID seen by Canadian paediatricians. The survey response rate was 27% (664/2490). Paediatricians reported 339 cases of ARFID in the previous 12 months.

The results identified that the majority of paediatricians (63%) were unfamiliar with ARFID and its specific diagnostic criteria. Thirty percent (30%, 239/657) of paediatricians who suspected a diagnosis of ARFID misdiagnosed due to inappropriate application of the exclusion criteria.

Canadian paediatricians are considerably unfamiliar with ARFID as a new diagnostic category, its diagnostic criteria, and clinical features. Although little is known about effective treatment for ARFID, experience to date suggests that children and adolescents with this condition require medical monitoring as well as nutritional and psychological interventions. Increased knowledge and awareness of ARFID will ultimately facilitate early recognition, accurate diagnosis, and immediate treatment of children and adolescents with this condition. Dissemination of information about ARFID will help to ensure that children and youth are adequately screened, assessed, and treated.

### Conclusion

- Education and dissemination of information about ARFID will increase awareness of this new disorder and ensure that children and youth are adequately screened, assessed, and treated.

### References

Available upon request from the CPSP office

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# Exposure to liquid detergent capsules

August 2014

MT Do, M Herbert, M Laffin Thibodeau, J Maguire

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### Highlights

- A one-time CPSP survey revealed 54 cases of children who were injured following exposure to liquid detergent packets in the past year.
- Forty-seven (87%) were injured following ingestion or exposure to the alimentary tract.
- Twenty-five (46%) of the injured children were admitted to hospital including six who were treated in intensive care units.

### Results

Since the 2012 introduction of single-load liquid detergent packets in North America, there has been a steady rise in child injuries associated with exposure to these products. A recent report from the United States Poison Control Centers reported that more than 17,000 children were exposed to laundry detergent packets over a two-year period, causing several hundred serious injuries and one death. The physical characteristics of liquid detergent packets – malleable, shiny, and brightly coloured – make them appealing to young children who mistake them for toys or candy.

A one-time survey through the CPSP was conducted to obtain more detailed information on child injuries following exposure to detergent packets in Canada. The survey was distributed to 2474 paediatricians and subspecialists. Thirty-seven of the 743 respondents (30% overall response rate) had treated a total of 54 children injured following exposure to liquid detergent packets in the past year.

Among the 54 exposed children, 30 (56%) were younger than 2 years of age, 23 (43%) were 2 to 4 years of age, and 1 was 5 years of age or older. All but two of the children were treated in emergency departments or inpatient settings. Forty-seven (87%) children were injured following ingestion or exposure to the alimentary tract. Six (11%) children sustained ocular injuries, 4 (7%) had dermal injuries, and 3 (6%) had airway exposure with pulmonary toxicity. Six children had multiple injuries. Adverse events included: nausea and vomiting (17); chemical burns to the mouth, esophagus or skin (15); pneumonitis (10); conjunctivitis (4); central nervous system depression (3); and corneal injuries (2). Twenty-five (46%) of the injured children were admitted to hospital including six who were treated in intensive care units. Six children (11%) were discharged from hospital with referral for medical follow-up and 21 (39%) children were discharged without need for follow-up (two cases were unspecified).

Forty (74%) cases involved exposure to detergent packets for laundry and 12 (22%) for dishwashers. Among the 34 incidents in which the point of access to the packets was known, two thirds (23) of the children took detergent packets directly from the manufacturer's original package or container and seven found a packet on the floor or lying around.

Exposure to liquid detergent packets can cause serious injury to children. Almost half of the children in this study required hospitalization with 11% having injuries so severe they required treatment in intensive care units. The results of this survey highlight the importance of educating parents and caregivers about the risks liquid detergent packets pose for young children, the safe storage and use of these detergent packets, and seeking medical attention should exposure occur.

### Conclusion

- Where point of access to the packets was known, two thirds of children took packets directly from the manufacturer's original package.
- Results of the survey highlight the serious injuries that can occur and the importance of educating parents and caregivers about the risks to young children.

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#### Principal investigator

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## RESEARCH OPPORTUNITIES

# Call for New Studies

### Wanted

Investigators to initiate new CPSP studies

### The program

- Well established, timely and cost-effective
- Multi-faceted surveillance, capable of collecting reliable data in a variety of fields
- Effective at monitoring low-frequency, high-impact diseases and conditions

### Track record

- 80% response from approximately 2500 paediatricians
- 87% data completion rate for identified cases

### Study ideas

- Bronchiectasis – non-cystic fibrosis
- Congenital syphilis
- End-stage renal disease in early infancy
- Gender dysphoria
- Health issues among refugee children in Canada
- Juvenile-onset recurrent respiratory papillomatosis
- Late-diagnosed cyanotic congenital heart disease
- Medical marijuana use in children
- Paediatric HIV
- Pet-related *Salmonella* infections
- Severe neonatal hypernatremia
- Severe sports-related head trauma
- Toxic shock syndrome

If you are interested in these or other studies, or for more program information, please contact 613-526-9397 ext. 239 or e-mail [cpsp@cps.ca](mailto:cpsp@cps.ca).



*"For close to 20 years, the CPSP has been an important collaborative asset for research, health policy development and the active surveillance of less common paediatric disorders. The hundreds of Canadian paediatricians and paediatric subspecialists who participate monthly in the program ensure that the CPSP is an effective way to foster continuing medical education on a wide spectrum of clinical conditions that might otherwise go largely unrecognized."*

Bryce Larke, MD, Professor of Pediatrics, University of Alberta, Edmonton, Alberta, 1975-2001; Chief Medical Officer of Health, Whitehorse, Yukon, 2001-2008; CPSP Steering Committee member, 2004-2010







**For more information on the  
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