



## CPSP 2014 results: What have we learned?

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The Canadian Paediatric Surveillance Program (CPSP), a joint project of the Canadian Paediatric Society and the Public Health Agency of Canada, has completed >50 studies investigating a variety of rare diseases and conditions. The year 2014 was once again successful, resulting in an overall response rate of 80% of paediatricians surveyed monthly and a completion rate of 87% for clinical detailed questionnaires for reported cases. The following are the main highlights from the 11 studies that were active or concluded in 2014 and two one-time surveys.

### FINAL STUDY RESULTS

#### Early-onset major depressive disorder

- Twenty-two cases of early-onset major depressive disorder were confirmed, including 86% impaired in  $\geq 3$  functional domains and 82% having  $\geq 1$  psychiatric comorbidity.
- Children with early-onset major depressive disorder experienced severe symptoms: 73% reported suicidal thoughts and 23% had attempted suicide.
- Parental history of a mood disorder, most commonly major depressive disorder, was present in 68% of cases.

#### Fragile X syndrome

- Thirty cases of fragile X syndrome were confirmed during the study period, and the average age at postnatal diagnosis was 3.5 years.
- A family history of fragile X syndrome or a fragile X-related condition was present in a minority (13%) of cases, of which four, ranging from 5 to 9 years of age, had a delayed diagnosis.
- Comorbid diagnoses included speech and communication problems, and autism.

#### Kawasaki disease

- The study confirmed 285 cases of Kawasaki disease.
- Most patients (93%) received intravenous immunoglobulin therapy; of these, 25% failed initial therapy and 11% developed evidence of intravenous immunoglobulin-associated hemolysis.
- Most patients recovered without cardiac sequelae, with 5% developing coronary artery aneurysm and 23% developing dilatation sometime during their disease course.

#### Periodic fever syndromes

- There were 179 confirmed cases of periodic fever syndromes.
- Periodic fever, aphthous stomatitis, pharyngitis and adenitis (PFAPA) syndrome was the most frequently reported diagnosis, followed by undefined periodic fever syndromes and familial Mediterranean fever.

- On average, symptom onset occurred 2.5 years before diagnosis and patients saw multiple physicians before diagnosis.

### ONGOING STUDY RESULTS

#### Acute flaccid paralysis

- Canada was declared polio-free in 1994; however, in accordance with WHO recommendations, Canada conducts acute flaccid paralysis (AFP) surveillance in the population <15 years of age to monitor for poliomyelitis due to ongoing transmission of wild poliovirus in other countries around the world.
- Thirty-seven cases of AFP were identified in 2014, representing a nonpolio AFP detection rate of 0.65 cases per 100,000 for children <15 years of age, which was below the rate of 1.0 case per 100,000 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 and transverse myelitis.
- The AFP surveillance protocol and detailed questionnaire have been updated, and a user manual was developed with the aim of improving Canada's ability to meet the WHO surveillance performance objectives.

#### Adverse drug reactions – serious and life-threatening

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

#### Childhood Lyme disease

- 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 one to 16 years.
- Of the 17 confirmed or probable cases, 10 were diagnosed in Nova Scotia and the remainder 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.

#### Childhood tuberculosis

- Sixty-three confirmed cases of childhood tuberculosis (TB) were identified in 2014. Information on ethnicity was available for

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51 Canadian-born children, the majority of whom were Aboriginal (n=35) and 12 were born overseas.

- The incidence of both multidrug-resistant TB and extensively drug-resistant TB is increasing in many parts of the world and is present in Canada.
- One case of multidrug-resistant TB was reported in a Canadian child in 2014.

#### **Hypoglycemia in low-risk term newborns**

- 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 6 h of life. Those who had hypoglycemia recognized beyond 6 h after birth had higher morbidity.
- A brain magnetic resonance image was obtained in 29% of cases; 71% of these exhibited evidence of hypoglycemic brain injury.
- Five cases had seizure at the time of presentation. One case presented abnormal neurological signs at the time of hospital discharge.

#### **Severe alcohol intoxication in adolescents**

- In the second year of this two-year surveillance study, 15 cases of severe alcohol intoxication in Canadian adolescents were identified.
- 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 death was reported in hospital.

#### **Sudden unexpected death in epilepsy**

- 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 the frequency of SUDEP is extremely low or Canadian paediatricians are not aware of such events occurring among children within the community.

### **ONE-TIME SURVEY RESULTS**

#### **Avoidant/restrictive food intake disorder**

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

#### **Exposure to liquid detergent capsules**

- The CPSP one-time 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.

The CPSP continues to undertake new studies and surveys investigating rare conditions or severe outcomes from common conditions, and this information serves to improve the quality of the day-to-day lives and care of Canadian children and youth. Future studies hold even more potential. This active epidemiological surveillance program enables large-scale collaboration that would not otherwise be possible with individual institution-based studies, and provides the framework for future prospective research, which can further define the overall findings of these surveillance studies.

Sincere thanks to all those who help make the CPSP work: the members of the Steering Committee who oversee the program and provide wise guidance; the investigators who initiate new proposals, bring them to fruition and publish their results; and, most importantly, the 2500 paediatricians and paediatric subspecialists who voluntarily respond each month.

For more information on the above studies, visit [www.cpsp.cps.ca/publications](http://www.cpsp.cps.ca/publications), as well as [www.cpsp.cps.ca/surveillance/current-studies](http://www.cpsp.cps.ca/surveillance/current-studies) and [www.cpsp.cps.ca/surveillance/concluded-studies](http://www.cpsp.cps.ca/surveillance/concluded-studies).

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*The Canadian Paediatric Surveillance Program (CPSP) is a joint project of the Canadian Paediatric Society and the Public Health Agency of Canada, which undertakes the surveillance of rare diseases and conditions in children and youth. For more information, visit our website at [www.cpsp.cps.ca](http://www.cpsp.cps.ca).*