

Canadian Paediatric Surveillance Program 2008 Quiz

The purpose of this quiz is to disseminate information gathered through the Canadian Paediatric Surveillance Program studies to guide physicians in their clinical practice.

1. The World Health Organization refers to public health surveillance as “Information for Action”. Describe how the Canadian Paediatric Surveillance Program helps achieve this objective.
2. What are the health hazards related to magnetic toys?
3. What are the most common presenting symptoms in juvenile idiopathic arthritis?
4. What are the most important factors affecting prognosis in infants with severe combined immunodeficiency?
5. Name a public health intervention that has favourably decreased the rate of neural tube defects.
6. What is the leading nongenetic cause of hearing impairment in children?
7. Are legislative measures for the prevention of lap-belt syndrome advancing in Canada?
8. Why is surveillance of adverse drug reactions important in paediatrics?
9. How does Prader-Willi syndrome occur?
10. What is neuroleptic malignant syndrome?

*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 Web site at <www.cps.ca/cpsp>.*

Answer key on page 849

Canadian Paediatric Surveillance Program 2008 quiz – Answers

Questions on page 842

1. The Canadian Paediatric Surveillance Program (CPSP) ensures that data providers receive timely feedback of study results to guide their clinical practice. This is achieved through different formats, such as publications of annual results, regular highlights in *Paediatrics & Child Health*, oral and poster presentations, and a regularly updated Web site. The Program also disseminates “Information for Action” to all relevant stakeholders to support the development of medical and public health interventions. National high-quality epidemiological evidence contributes to improved health for children and youth.
2. Toys containing 3 mm to 8 mm magnets carry the risk of ingestion or aspiration. If more than one is swallowed, they can attract each other in different segments of the bowel causing serious injury and requiring surgical removal. The CPSP 2007 one-time survey identified 20 cases, including children with small bowel obstruction and perforation. Parents and other caregivers should ensure that magnetic toys are not kept in the surroundings of children younger than six years of age.
3. Chronic arthritis in children and adolescents, called juvenile idiopathic arthritis (JIA), is a long-term disorder that can occur at any age. In the first three months of surveillance, 117 cases of newly diagnosed JIA were confirmed. In these cases, the most common presenting symptoms were joint pain, swelling or morning stiffness. It is important to note that the absence of joint pain and joint swelling was reported in 10% and 15% of patients, respectively. Often, the lack of joint pain is a factor leading to delayed diagnosis with the potential risk of joint damage and disability.
4. Severe combined immunodeficiency (SCID) is a group of genetic disorders characterized by profound abnormalities in T and B cells and natural killer cell development and function. SCID is associated with serious life-threatening infections with high morbidity and mortality. The most important prognostic factors are the early confirmation of the diagnosis and the performance of a bone marrow transplant before the appearance of overwhelming infections.
5. Folic acid fortification of cereal grain products is a public health intervention, mandated in 1998, that has favourably decreased the Canadian prevalence of neural tube defects (NTDs) from 1.58 to 0.86 per 1000 births. This primary prevention measure is important, because the 1997 to 1998 CPSP NTD study revealed that 80.6% of women with a pregnancy affected by an NTD, for whom information was available, had not taken folic acid before and during their pregnancies. This occurred in spite of current medical literature attesting that periconceptional use of folic acid decreases the risk of NTDs and other possible non-NTD birth defects, such as congenital cardiac and genitourinary defects, cleft palates and limb reduction deficits.

Continued on page 856

Canadian Paediatric Surveillance Program 2008 quiz – Answers

Continued from page 849

6. Congenital cytomegalovirus (CMV) infection is the leading nongenetic cause of hearing impairment in children. Even if asymptomatic at birth, 5% to 17% of infants with congenital CMV infection will have neurodevelopmental abnormalities, including progressive sensorineural hearing loss. The CPSP CMV study, from March 2005 to February 2008, revealed that, of the 35 of 48 cases with test results, nine cases had abnormal hearing assessment before discharge. Symptomatic or at-risk infants should have a hearing screening done before discharge from hospital and then close audiology follow-up. Early diagnosis and effective early interventions are essential to improve outcomes in hearing-impaired children.
7. Yes, but there is more work to be done. The 2005 CPS status report "Are We Doing Enough?" stated that only Ontario had appropriate legislation. By July 2008, only six provinces and no territories had legislated on booster seat use. The lap-belt syndrome (LBS) surveillance study, from September 2003 to August 2005, documented seven of 28 (25%) cases with permanent spinal cord lesions and 24 of 28 (85%) cases with abdominal injuries. To prevent LBS, the CPS recommends longer use of booster seats in children younger than eight years of age, weighing 18 kg to 36 kg and/or measuring less than 148 cm. More advocacy efforts are needed to ensure that the remaining provinces and territories will legislate and adopt reinforcement measures. Paediatricians should be aware of this syndrome and use every contact with families to promote the use of booster seats.
8. Adverse drug reactions (ADRs) represent a major cause of childhood morbidity and mortality. In 2007, 41 cases of suspected serious and life-threatening paediatric ADRs were confirmed by the surveillance study. Postmarketing surveillance is essential because premarket clinical drug trials often do not include children, and many drug products are not labelled for use in specific paediatric age groups. Postmarketing ADR surveillance can detect signals and provide data on a drug safety profile, which are critical in improving care in the paediatric population.
9. Prader-Willi syndrome (PWS) is a very rare genetic disorder, due to an abnormality on chromosome 15 with a paternal interstitial deletion in 70% of patients. Other less common mechanisms include uniparental disomy (approximately 28%), sporadic mutations, chromosome translocations and gene deletions. Children with PWS have difficulty in controlling their hunger and in sensing satiety, which may result in aggressive food-seeking behaviour, food foraging, hoarding and overeating, with obesity and its complications. A multidisciplinary approach with an energy-controlled dietary management program should alleviate the problems of overeating and prevent medical complications of obesity, such as diabetes, osteoporosis and heart failure.
10. Neuroleptic malignant syndrome (NMS) is a rare event associated with neuroleptic drugs, such as risperidone, fluoxetine and haloperidol. A higher dosage, or rapid and large increases, can trigger the development of NMS, which is potentially fatal in 25% of cases. The pathophysiology is thought to be related to decreased levels of dopamine either as a blockade or a genetically-reduced function of dopamine receptors. Clinical presentation includes high fever, profuse sweating, tachyarrhythmia, stiffness of extremities, marked elevation of creatinine phosphokinase and myoglobinuria. Physicians should stop neuroleptic drugs at the earliest signs and symptoms of this syndrome, and administer intravenous sodium dantrolene.