Can surveillance provide epidemiological data on Aboriginal health?

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The Canadian Paediatric Surveillance Program (CPSP) was established in 1996. This pan-Canadian active surveillance network of more than 2500 practising clinical paediatricians and paediatric subspecialists collects timely epidemiological data on rare paediatric diseases and conditions. In 2004, the Program was favourably evaluated by an external expert advisory group (EAG) (1). One of the recommendations of the EAG was for the CPSP to capture the unique entity of northern Canada by undertaking surveillance studies on diseases affecting the health of the Aboriginal population.

Aboriginal (First Nations, Métis and Inuit) children and youth represent 5.8% of the entire Canadian paediatric population. Certain diseases, some with serious and even fatal outcomes, seem to have a higher incidence in the Aboriginal population. However, for many of these diseases, there is a lack of national frequency data. Epidemiological and clinical information are essential to document the burden of illness, guide clinical care and support public health actions.

This article presents national epidemiological data, where information and descriptive analysis were available, on six studies undertaken by the CPSP with data on Aboriginal children: severe combined immunodeficiency diseases (SCID), neonatal herpes simplex virus infection (NHSV), congenital cytomegalovirus infection (CMV), juvenile idiopathic arthritis (JIA), non-type 1 diabetes mellitus (NT1DM) and persistent albuminuria in type 2 diabetes mellitus (AT2D).

METHODS

Study proposals were reviewed and approved by the CPSP Steering Committee, a multidisciplinary group of experts that includes the Canadian Paediatric Society, the Public Health Agency of Canada, the Paediatric Chairs of Canada, and liaison representatives from various subspecialties and the Canadian Immunization Monitoring Program – ACTive (IMPACT). After approval by the principal investigators' local Research Ethics Board, the studies were included in the Program for a minimum period of two years. CPSP participants received a monthly reporting form listing the current conditions under surveillance. For each condition, respondents indicated the number of new cases seen in the last month, including 'nil' reports. For each case reported, a follow-up, detailed, clinical questionnaire was completed. The national average report rates for the initial report and the detailed questionnaire are 80% and 87%, respectively.

RESULTS

Severe combined immunodeficiency (SCID) – April 2004 to March 2010

SCID includes several rare life-threatening genetic diseases with high morbidity and mortality. The general SCID incidence is one case in 75,000 to 100,000 live births (2). Although the incidence rate may vary from country to country, it was found to be much higher in the US Navajo population (52 in 100,000 live births). The Canadian incidence was unknown as no data were available. The SCID study confirmed 44 cases, representing an incidence of two cases in 100,000 live births. While Aboriginal children accounted for 16% of the confirmed cases, they accounted for only 5.6% of the children under the age of five in Canada in 2006. The average age at diagnosis for all confirmed cases was four months. This study reinforced the importance of earlier diagnosis, which carries a better prognosis, as hematopoietic stem cell transplantation can be performed before the appearance of opportunistic infections (3).

Neonatal herpes simplex virus infection (NHSV) – October 2000 to September 2003

Herpes simplex virus infections pose a public health concern, especially since a high proportion of these infections can be asymptomatic. One of the most serious consequences of genital HSV is the perinatal maternal-child transmission. This CPSP surveillance study provided the first Canadian national data for 58 confirmed cases of NHSV infections over three years. The estimated incidence was 5.9 cases per 100,000 live births, with a prematurity rate of 28% and a fatality rate of 16%. Aboriginal women/infants appear disproportionately affected (10.5%). The majority of typed cases (62.5%) were HSV-1, which has implications for future vaccine development (4,5).

Congenital cytomegalovirus infection (CMV) – March 2005 to February 2008

Congenital CMV infection is the most common congenital infection, affecting from 0.3% to 2.2% of all live births (6,7), but Canadian data are scarce. This study confirmed 49 cases of congenital CMV, representing 0.54 cases per 10,000 live births. New Canadian, First Nations and rural children appeared to be at higher risk; 67% of the infected infants born in rural Canada were of First Nations origin (four of six cases). First Nations women/infants appear disproportionately affected (13%). Congenital CMV caused significant morbidity during the neonatal reporting period, with affected infants experiencing prolonged hospital stays of high intensity. Median length of stay reported

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was 25 days with a median of 10 days in the intensive care unit. Four infants died, for an early mortality rate of 8%. At the time of the study, only severely affected infants in the prenatal or neonatal period were being detected by paediatricians; of note, only one-half of the infants with neurological disease were being treated with antiviral therapy. Many infected infants presenting either without symptoms or with milder symptoms were not identified and offered appropriate therapy and follow-up. Congenital CMV is the most common cause of non-hereditary deafness in children and may be progressive over the years. In order to optimize outcomes for these infants, routine screening should be considered as a public health policy (8).

Juvenile idiopathic arthritis (JIA) – October 2007 to September 2009

Juvenile idiopathic arthritis is a rare condition that can result in serious long-term disability in children and adolescents. Accurate Canadian data on the scope of chronic arthritis in children are scarce. This two-year surveillance study confirmed 846 cases, resulting in an incidence of 4.3 new cases per 100,000 children. The study data demonstrated that JIA was seen across Canada, and it documented cases in Aboriginal children from the Northwest Territories, Yukon and Nunavut. First Nations and Inuit children represented 4% of the confirmed cases. The most common form of JIA was the oligoarticular subtype. The median time from symptom onset to diagnosis was 4.3 months, with the involvement of multiple health care providers. The study also demonstrated that to gain early disease control, current treatment included a nonsteroidal anti-inflammatory medication, intra-articular steroid injections and a disease modifying agent (9).

Non-type 1 diabetes mellitus (NT1DM) – April 2006 to March 2008

With the rapidly increasing prevalence of childhood and youth obesity, type 2 diabetes (T2D) is also increasing. There is a global effort to conduct epidemiological studies to quantify the extent of the problem. As data on the incidence and prevalence of T2D in Canadian children were limited, the CPSP study was conducted and confirmed 345 cases of NT1DM (ie, type 2 diabetes mellitus (T2DM), monogenic forms of diabetes and secondary diabetes including medication-induced diabetes mellitus), including 227 cases of T2D. Of these, 100 (44.1%) cases were Aboriginal. Amongst the children with T2D, 95% were obese and nearly 40% had at least one obesity-related comorbid condition at diagnosis. The observed minimum incidence rate of T2D in Canadian children <18 years of age was 1.54 per 100,000 per year with the Canadian Aboriginal children having the highest incidence at 23.2 cases per 100,000 per year. This is comparable to the incidence of T2D in the US Navajo paediatric population (10). Study results identified the need for national randomized control trials of efficacy and safety of various treatment modalities for T2D in the paediatric population (11,12).

Persistent albuminuria in the paediatric population with type 2 diabetes mellitus (AT2D) – April 2010 to March 2012

The natural history of T2D in childhood is largely unknown. Evidence suggests that complications occur at an earlier age with a shorter duration of diabetes in childhood-onset T2D. The first sign of diabetic nephropathy is persistent microalbuminuria, which may progress to end-stage renal failure. After 21 months of surveillance, 30 cases of persistent albuminuria were confirmed, and children of Aboriginal heritage appear to be overwhelmingly affected (28/30, 93%). The majority of Aboriginal children reported are of First Nations heritage (26/28). The mean age at diagnosis of albuminuria was 13.6 years (range 9.8 to 17.7 years). National epidemiological data is important to define the spectrum and the extent of the problem, to predict burden of illness and to plan screening and intervention programs (13).

DISCUSSION

An active surveillance network well connected with front-line practising paediatricians and public health officials can provide valuable national data on childhood disorders despite their low frequency. However, no population-based surveillance study will achieve complete ascertainment. Researchers encounter limiting factors such as, some paediatricians are nonparticipants or nonresponders; other paediatricians might omit to report thinking that a colleague had already reported the case; children might have milder forms of disease and not be recognized; children living in geographically isolated or remote regions of Canada where few paediatricians are practising are often treated by nonparticipating health care providers. Nonetheless, for the majority of diseases included in the CPSP, the severity is such that most northern children will be transferred to southern university paediatric health centres for their treatment where paediatricians and paediatric subspecialists actively participate in the Program.

CONCLUSION

The surveillance studies described in this article illustrate the importance of collecting national data to have a better comprehension of these rare childhood diseases and to better inform prevention strategies. All have captured cases in the Aboriginal population, often documenting an over-representation. Further studies with added collaborators from remote regions and from northern nursing stations would improve case ascertainment. In 2011, the CPSP initiated a multiphase online reporting process that might provide new opportunities for improving the collection of data from northern communities that have Internet access.

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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.cps.ca/cpsp.