CANADIAN PAEDIATRIC SURVEILLANCE PROGRAM

2003 RESULTS

Canadian Paediatric Society
Société canadienne de pédiatrie

Health Canada
Santé Canada

Mission Statement

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.

For more information on the Canadian Paediatric Surveillance Program or a French version of this report, please contact:

Canadian Paediatric Society

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Acknowledgements

The key strengths of the CPSP continue to be the participation of Canadian paediatricians, subspecialists and other health-care providers in the monthly collection of information on rare paediatric conditions, our principal investigators who review and analyze the data collected to provide us with knowledge and educational solutions to help children and youth around the world, and our Steering Committee members who continue to guide the program.

For their role in the verification of data collected, we thank:

- Canadian Association of Paediatric Health Centres
- Canadian Institute for Health Information
- Canadian Paediatric Decision Support Network
- IMPACT (Immunization Monitoring Program ACTive) centres
- Notifiable Diseases Reporting System, Centre for Infectious Disease Prevention and Control, Health Canada

We also gratefully acknowledge the financial support received to maintain and expand the program in order to improve the health of Canadian children and youth. A summary of supporters is found in this report (page 8).

The strong CPSP partnership between the Canadian Paediatric Society (CPS) and Health Canada's Centre for Infectious Disease Prevention and Control (CIDPC) allows the program to grow in Canada and to take a leadership role on the international scene.

A special thank you

To all who participated in the Canadian Paediatric Surveillance Program's external evaluation, thank you. With your cooperation and involvement, it was possible to provide the Expert Advisory Group (EAG) with valuable information with which to assess the merits of the program. A special thank you is extended to the EAG, chaired by Dr. Robert McMurtry, for evaluating the program to determine its strengths and weaknesses and how well it is achieving its objectives and goals.

Dr. Robert McMurtry
"CPSP, a gem, a light under a bushel"

The Steering Committee is proud of the results of the review, which stated, "The CPSP represents an important collaborative tool for surveillance, research and policy development. In this role, it was perceived as unique in Canada. In other words, it provides an important activity that would disappear in its absence, unless a much larger investment was made to replace it."

You are encouraged to read the executive summary of the evaluation results on page 14 of this report. A copy of the complete evaluation report is available through the CPSP Senior Coordinator.

Congratulations all!

Foreword

Federal Minister of Health, Health Canada


The Honourable Pierre S. Pettigrew

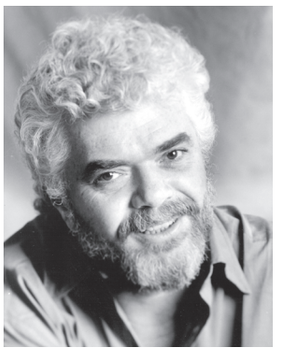
As Minister of Health, I wish to congratulate the Canadian Paediatric Society on the successful completion of the eighth year of the Canadian Paediatric Surveillance Program (CPSP). It is well recognized that Canadian paediatricians play a vital role in the program.

The knowledge generated by the CPSP enables paediatricians and the health-care community to educate other health-care professionals, improve the quality of life for children with rare conditions, and increase public awareness, both in Canada and internationally. Thank you to the 2,300 paediatricians who regularly take the time to return the monthly report.

Canadian society values its children. It is important that we foster and nurture their mental and physical well-being. In partnership, Health Canada and the Canadian Paediatric Society will continue to work towards the goal of improving the health and well-being of Canada's children.

I wish the Canadian Paediatric Society many years of success with the Canadian Paediatric Surveillance Program.

Director General, Centre for Infectious Disease Prevention and Control


Dr. Frank A. Plummer

I am pleased to accept the eighth annual report of the Canadian Paediatric Surveillance Program (CPSP). I am heartened by the success of the program, especially CPSP's leadership in strengthening the International Network of Paediatric Surveillance Units where the health of children exemplifies a 'global village'.

I would like to reaffirm my commitment to the partnership between the Centre for Infectious Disease Prevention and Control (CIDPC) and the Canadian Paediatric Society (CPS). An Expert Advisory Group (EAG), convened to oversee the CPSP evaluation, emphasized the importance of the relationship by stating that "an important collaboration has been established between the CPS and Health Canada which is exemplar". The EAG also concluded that the CPSP is an invaluable tool for future expanded surveillance and/or specific outbreaks because the program's network of over 2,300 paediatricians, coupled with centralized coordination by the CPS, strategically enables it to rapidly respond to public health emergencies.

I would like to personally thank the CIDPC staff who have played, and will continue to play, a major role in the success of the program by serving in multiple capacities as program managers, members of the CPSP Steering Committee and/or principal investigators. The CIDPC continues to financially support the infrastructure of the program and to coordinate funding for several Health Canada studies. Departmental studies undertaken in 2003 include monitoring of acute flaccid paralysis, congenital rubella syndrome, neonatal herpes simplex virus infection, and early-onset eating disorders. Future and proposed departmental studies include paediatric serious and life-threatening adverse drug reactions and severe combined immunodeficiency syndrome.

My gratitude would not be complete without attributing the success of the program to the paediatricians of Canada who diligently complete the monthly check-off form and who provide detailed information for each reported case.

President of the Canadian Paediatric Society


Dr. Diane Sacks

Congratulations to us: They asked – We answered

If there was any doubt that Canadian paediatricians care and are interested in all matters that affect children and youth, look no further than our response to the CPSP.

On a regular basis, I get the monthly forms in my office from the CPSP. I must admit that some of the conditions I remember only vaguely from my medical school days, long, long ago. Yet, every month I fill out the form and send it back, knowing it is important to learn more about these conditions and, in some cases, to find out that they are not in fact so rare. And so it appears do many, many of my colleagues from all over the country because the response rate in 2003 was an astounding 83%. The voluntary questionnaire completion rate was an unbelievable 96%, and 67 new study suggestions were forthcoming for further evaluation. These are truly remarkable rates, as anyone who has ever done any surveying can tell you. In fact, that is just what the Expert Advisory Group, led by Dr. Robert McMurtry, noted in their external evaluation when they praised the program. The surveillance project has raised awareness of the work of the Canadian Paediatric Society and paediatricians within our own government circles and internationally.

Congratulations to those who run the program, and to us. Well done!

CPSP Chairman


Dr. Gilles Delage

What a great year! Not only did the CPSP continue to reinforce the national collaborative network of active surveillance to provide reliable data on high-impact, low-frequency conditions, but at the same time, the program also underwent a very thorough external evaluation.

In the previous report, I mentioned that this formal program evaluation was being undertaken to answer important questions concerning the program's strengths, weaknesses and future course of action. To this end, an international Expert Advisory Group was formed to review the program's policies, objectives, targets, strategies, action plans, performance and output. I am pleased to report that after an extensive review, the program passed with flying colours. The group was unanimous in their opinion that the CPSP "represents excellent value for money; the achievement in this respect was seen as excellent and unsurpassed by any comparable program known to the Expert Advisory Group".

As Steering Committee Chair, I am deeply grateful to Dr. Robert McMurtry and the members of the advisory group for their insight into the strengths of the CPSP as an active surveillance tool, their guidance in areas of future expansion and their vision of the CPSP's worth in the paediatric and public health research communities. I encourage you to read their report on page 14.

We are proud of the CPSP's achievements to date and are committed to enhancing its activities to continue making a major contribution to the health of Canadian children, by influencing paediatric practice and public health policy.

CPSP Steering Committee

Dr. Gilles Delage (Chair)	Canadian Paediatric Society
Dr. Garth Bruce	Canadian Paediatric Society
Dr. Rick Cooper	Paediatric Chairs of Canada
Ms. Marie Adèle Davis	Canadian Paediatric Society
Ms. Jo-Anne Doherty	Centre for Infectious Disease Prevention and Control, Health Canada
Dr. Danielle Grenier	Canadian Paediatric Society
Dr. Richard Haber	Canadian Paediatric Society
Dr. Susan King	Canadian Paediatric Society
Dr. Simon Levin	Canadian Association of Child Neurology (Liaison)
Dr. Catherine McCourt	Centre for Healthy Human Development, Health Canada
Ms. Andrea Medaglia	Canadian Paediatric Society
Mr. Paul Muirhead	Consultant
Dr. Jeff Scott	Council of Chief Medical Officers of Health
Dr. Anne Summers	Canadian College of Medical Geneticists (Liaison)
Dr. Paul Varughese	Centre for Infectious Disease Prevention and Control, Health Canada
Dr. Wendy Vaudry	IMPACT (Immunization Monitoring Program ACTive)
Dr. Lynne Warda	Canadian Paediatric Society
Dr. Lonnie Zwaigenbaum	Canadian Paediatric Society

CPSP Working Group

Ms. Andrea Medaglia (Chair)	Canadian Paediatric Society
Ms. Marie Adèle Davis	Canadian Paediatric Society
Ms. Jo-Anne Doherty	Centre for Infectious Disease Prevention and Control, Health Canada
Dr. Danielle Grenier	Canadian Paediatric Society

Publications in 2003

Published papers related to studies

(See http://www.cps.ca/english/cpsp for a complete list of abstracts with hotlinks.)

Paediatric adverse drug reactions can be fatal. Grenier D, Doherty J, Medaglia A. *Paediatr Child Health* 2003;8(4):218

Carrier frequency of the Smith-Lemli-Opitz IVS8-1G>C mutation of the DHCR7 gene in African-Americans. Wright BS, Nwokoro NA, Waye JS, Wassif CW, Nowaczyk MJM, Porter FD. *Am J Med Genet* 2003 (published on-line January 17)

Though published in 2004, the following papers are noteworthy:

Canadian Paediatric Surveillance Program : A developmental check-up. Scott J. *Paediatr Child Health* 2004;9(1):13-4

Canadian Paediatric Surveillance Program confirms low incidence of hemorrhagic disease of the newborn in Canada. *Paediatr Child Health* 2004;9(4):235-8

Highlights published in *Paediatrics & Child Health*

(See http://www.cps.ca/english/cpsp for a complete list of highlights with hotlinks.)

Do you know the answers? – CPSP Quiz. *Paediatr Child Health* 2003;8(10):615,641

The unrealistic quest to thinness. *Paediatr Child Health* 2003;8(9):563

Helping to prevent obesity and complications in children with Prader-Willi syndrome. *Paediatr Child Health* 2003;8(8):510

CPSP 2002 Results: What have we learned? *Paediatr Child Health* 2003;8(7):447

Lap-belt injuries: A view from the bedside. *Paediatr Child Health* 2003;8(6):373

Survey on lap-belt syndrome: Results and next steps. *Paediatr Child Health* 2003;8(6):374

Call for new studies: Research opportunities. *Paediatr Child Health* 2003;8(5):297

How to improve your diagnostic tools for genetic disorders. *Paediatr Child Health* 2003;8(4):217

Is the risk of kernicterus rising? *Paediatr Child Health* 2003;8(3):150

Congenital rubella syndrome – Time to act on missed prevention opportunities. *Paediatr Child Health* 2003;8(2):107-8

Surveillance of rare genetic disorders: No longer orphan diseases. *Paediatr Child Health* 2003;8(1):55

Presentations in 2003

(See http://www.cps.ca/english/cpsp for a complete list of presentations with hotlinks.)

National

Vitamin D deficiency rickets among children living in Canada: A new look at an old disease. Ward LM, Gaboury I, Ladhani M, Zlotkin S. Presented at the CDA/CSEM Professional Conference, Ottawa, October 2003.

Risk factors for vitamin D deficiency rickets among children living in Canada: Results of an incidence study through the Canadian Paediatric Surveillance Program. Ward LM, Gaboury I, Ladhani M, Zlotkin S. Presented at the Canadian Society of Endocrinology and Metabolism's Annual Meeting, Ottawa, October 2003.

Puberty and growth in CHARGE association/syndrome. Blake K. Presented at the 13th Dbl World Conference of Deafblindness, Mississauga, August 7, 2003.

Central nerves of CHARGE association/syndrome. Blake K. Presented at the 13th Dbl World Conference of Deafblindness, Mississauga, August 5 to 10, 2003.

CHARGE into the adolescent and adult decades. Blake K. Presented at the 13th Dbl World Conference of Deafblindness, Mississauga, August 5 to 10, 2003.

Neonatal herpes simplex virus infections in Canada. Wong T, Burton S, Cormier L, Embree J, Steben M, Rusen ID. Presented at the International Society for Sexually Transmitted Disease Research Conference, Ottawa, July 27 to 30, 2003.

Surveillance helping patients with orphan genetic disorders. Summers A, Berall G, Blake KD, Nowaczyk MJM, Desantadina MV. Presented at the 80th Annual Meeting of the Canadian Paediatric Society, Calgary, June 21, 2003.

Public health implications of the Canadian Paediatric Surveillance Program. Grenier D, Doherty J, Medaglia A. Presented at the 80th Annual Meeting of the Canadian Paediatric Society, Calgary, June 19, 2003.

C.H.A.R.G.E. account: The health economics of managing CHARGE syndrome. Budden H, Blake KD. Presented at the 80th Annual Meeting of the Canadian Paediatric Society, Calgary, June 18 to 22, 2003.

The incidence and prevalence of CHARGE association/syndrome in Canada. Issekutz KA, Smith IM, Prasad C, Graham JM, Blake KD. Presented at the 80th Annual Meeting of the Canadian Paediatric Society, Calgary, June 18 to 22, 2003, and at the 13th DbI World Conference of Deafblindness, Mississauga, August 7, 2003.

The cranial nerve anomalies of CHARGE association/syndrome (A/S). Lawand CMD, Blake KD, Prasad C, Graham JM Jr. Presented at the 80th Annual Meeting of the Canadian Paediatric Society, Calgary, June 18 to 22, 2003.

International Network of Paediatric Surveillance Units: A child's global village. Grenier D, Doherty J, Medaglia A. Presented at Child & Youth Health Congress, Vancouver, May 14, 2003, and at the Irish and American Paediatric Society Meeting, Ottawa, September 20, 2003.

Oral sensory experiences and feeding issues in CHARGE syndrome. Marche DM, Dobbelsteyn C, Rashid M, Blake KD. Presented at the 28th Annual Conference of the Canadian Association of Speech-Language Pathologists and Audiologists, St. John's, May 2003.

International

Genetic studies: A significant component of the Canadian Paediatric Surveillance Program. Summers A. Presented at the American Society of Human Genetics Annual Meeting, Los Angeles, November 5, 2003.

Challenging behavioural problems in children with genetic and rare conditions: Role of the Canadian Paediatric Surveillance Program. Grenier D, Doherty J, Medaglia A. Presented at the Europaediatrics 2003 Congress, Prague, October 22, 2003.

Adolescenthood and CHARGE syndrome. Blake K. Presented to Project Directors, National Technical

Assistance Consortium – Office of Special Education Programs, Washington, October 2003.

Rare diseases research through surveillance: The Canadian experience. Grenier D, Doherty J, Medaglia A. Presented at the European Society of Paediatric Research Meeting, Bilbao, September 28, 2003.

Puberty in CHARGE. Blake K. Workshop presented at the 6th International CHARGE Syndrome Conference, Cleveland, July 25 to 27, 2003.

General endocrine issues in CHARGE. Blake K. Workshop presented at the 6th International CHARGE Syndrome Conference, Cleveland, July 25 to 27, 2003.

National study of paediatric hemolytic uremic syndrome in Canada. Sockett P, Proulx F. Presented at the 5th International Symposium on Shiga Toxin (Verocytotoxin)-Producing *Escherichia coli* Infections (VTEC 2003), Edinbugh, June 8 to 11, 2003.

Cerebral edema (CE) in pediatric diabetic ketoacidosis (DKA) in Canada. Cummings EA, Lawrence SE, Daneman D. *Diabetes* 2003; 52(Suppl1):A400. Presented at the 63rd Scientific Sessions of the American Diabetes Association, New Orleans, June 2003.

Funding

To date, funding for the surveillance program has been made available from the Centre for Infectious Disease Prevention and Control, Health Canada, as well as other government departments, organizations and companies interested in increased knowledge of uncommon childhood conditions and the practical improvement in prevention and treatment.

Funding is required for program management including administrative and financial support. Educational grants are welcome from all interested in monitoring and contributing to the improvement of health of Canadian children and youth.

We gratefully acknowledge funding from the following sources:

Government departments:
Health Canada
- Population and Public Health Branch
  - Centre for Healthy Human Development
    - Division of Childhood and Adolescence
    - Health Surveillance and Epidemiology Division
    - Healthy Communities Division
  - Centre for Infectious Disease Prevention and Control
    - Division of Community Acquired Infections
    - Division of Immunization and Respiratory Diseases
    - Division of Surveillance and Risk Assessment
- Health Products and Food Branch
  - Office of Nutrition Policy and Promotion
    - Food and Nutrition Surveillance

Transport Canada
- Safety and Security Group
  - Road Safety and Motor Vehicle Regulation

Non-governmental sources:
- CHARGE Syndrome Foundation, Inc.
- Dairy Farmers of Canada
- Fondation de la recherche sur les maladies du Québec
- GlaxoSmithKline
- IWK Health Centre
- Mead Johnson Nutritionals
- Merck Frosst Canada Ltd.
- North York General Hospital
- Ontario Prader-Willi Syndrome Association
- Striving for Excellence Fund, Mount Sinai Hospital
- The Physicians' Services Incorporated Foundation

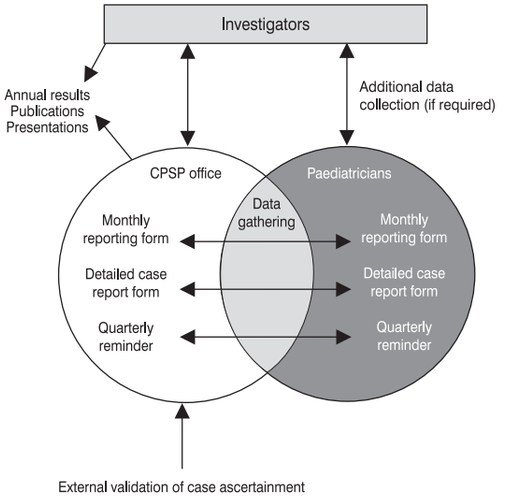
Surveillance at Work

Overview

Surveillance is an important part of the practice of medicine allowing for the tracking and studying of conditions. Not only can the burden of disease be determined and interventions to prevent the occurrence of a disorder be assessed, but information collected can also allow development of future health policies to address the needs of patients with these conditions. The CPSP is designed to study uncommon disorders with high morbidity and mortality in childhood or rare complications of more common diseases of such low frequency that data collection nationally is required to generate a sufficient number of cases to derive meaningful results. When the CPSP Steering Committee reviews new study proposals, preference is given to studies that have strong public health importance or could not be undertaken any other way. All studies must conform to high standards of scientific rigour and practicality.

FIGURE 1

Reporting process summary



Upon initiation of a new study, program participants receive a summary of the protocol, including the case definition and a brief description of the condition. In addition to providing a uniform basis for reporting, this approach serves to educate and increase awareness of low-frequency conditions with high morbidity and/or mortality.

The CPSP uses a two-tiered reporting process to ascertain and investigate cases: an initial 'check-off' form and a detailed reporting form. The full process is summarized in Figure 1.

Reporting

The initial reporting form, listing the conditions currently under surveillance, is mailed monthly to practising Canadian paediatricians and relevant paediatric subspecialists and health-care providers. Respondents are asked to indicate, against each condition, the number of new cases seen in the last month, including 'nil' reports. A 'nil' report is very important in active surveillance, as the CPSP cannot simply assume that no reply means no cases.

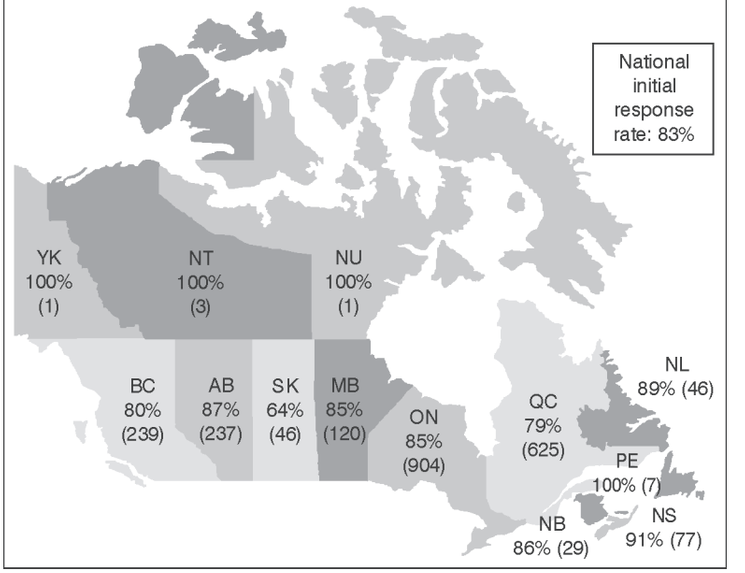
Participants report all cases meeting the case definitions, including suspect or probable cases where there is some doubt about reporting. This sometimes leads to duplicate reports but avoids missed cases. Case ascertainment is monitored and verified by investigating duplicate reports and comparing data with relative programs or centres.

The CPSP assures the confidentiality of all information provided to the program. Only non-nominal patient information, such as the date of birth, sex of the child and comments on the

condition, is requested for each reported case. This anonymous information is used to identify duplicates and is entered, as a reminder, on a detailed reporting form, which is sent to the original respondent to request case-specific information. Once the detailed report is returned to the CPSP, it is forwarded to the investigator for analysis. The investigator is responsible for contacting the respondent if further information is required. The CPSP is encouraged by the 96% response rate for completion of detailed questionnaires (see Table 1 for study breakdown).

FIGURE 2

Initial response rates (%) and number of participants for 2003



Respondents who do not reply every month receive quarterly reminders. As well, information, including the monthly compliance rates and the number of cases reported, is mailed quarterly to all participants to keep them informed of progress.

To thank paediatricians and paediatric subspecialists for their tremendous commitment to, and support of, the CPSP, 1,664 personal certificates were sent to acknowledge participation in 2003, and 360 letters of thanks went to participants who reported a case in 2003. An early-bird draw was

TABLE 1

2003 detailed questionnaire completion rates

Studies/conditions	Reported cases	Pending	% Completion rate
Acute flaccid paralysis	84	0	100
CHARGE association/syndrome	37	1	97
Congenital rubella syndrome	2	0	100
Early-onset eating disorders	93	0	100
Lap-belt syndrome	8	0	100
Necrotizing fasciitis	27	0	100
Neonatal herpes simplex virus infection	18	0	100
Neonatal hyperbilirubinemia – severe	178	6	97
Prader-Willi syndrome	56	7	88
Vitamin D deficiency rickets	80	7	91
Total number of cases (all studies)	**583**	**21**	**96**

held and complimentary copies of the AAP publication *Challenges in Pediatric Diagnosis* were awarded to Drs. Bryan Magwood (MB) and Claudio Fregonas (ON). Lucky winners in the year-end draw for one of two prizes to attend the June 2004 CPS Annual Conference in Montreal were Dr. Esias Van Rensburg (BC) who responded for all months in 2003, and Dr. Philip Mantynen (ON) who completed and returned a questionnaire for a reported case.

Participant workload

Program evaluation respondents indicated that the monthly reporting system is simple and 80% felt that the follow-up study questionnaires were easy to complete. As only non-nominal, non-identifiable data is collected through the CPSP, 90% of those who reported a case did not hesitate to provide clinical information. Even with a total of 583 reported cases in 2003, the majority of participants (1,975 of 2,335, 84.6%) had 'nil' cases to report. The importance of zero reporting must however be re-emphasized. As studies come and go, the workload shifts to different subspecialties. The number of reported cases was higher this year due to the neonatal hyperbilirubinemia and vitamin D deficiency rickets studies and the inclusion of the early-onset eating disorders study to the program.

Figure 3 illustrates the number of cases reported by respondents in 2003. It shows that most participants (84.6%) had no cases to report and checked off the 'nothing to report' box each month. In fact, 11.1% of participants reported one case and 3.2% reported two or three cases. Only 25 participants (1.1%) completed four or more questionnaires. It is interesting to note that 125 of the 583 reported cases were duplicates, validating CPSP ascertainment. The CPSP is extremely grateful that the majority of participants faithfully complete the detailed questionnaires subsequent to reporting cases. This demonstrates that they appreciate the enormous value of the scientific data collected. On their part, the Steering Committee continues to insist on short, precise and pertinent detailed questionnaires.

One-time survey questions

The CPSP is available as an inexpensive tool to survey participants on a one-time-basis in order to identify the prevalence of a problem or to answer a specific question. Once approved by the CPSP Steering Committee, the one-time survey question is sent to all participants with a monthly initial

FIGURE 3

Number of cases reported by respondents in 2003

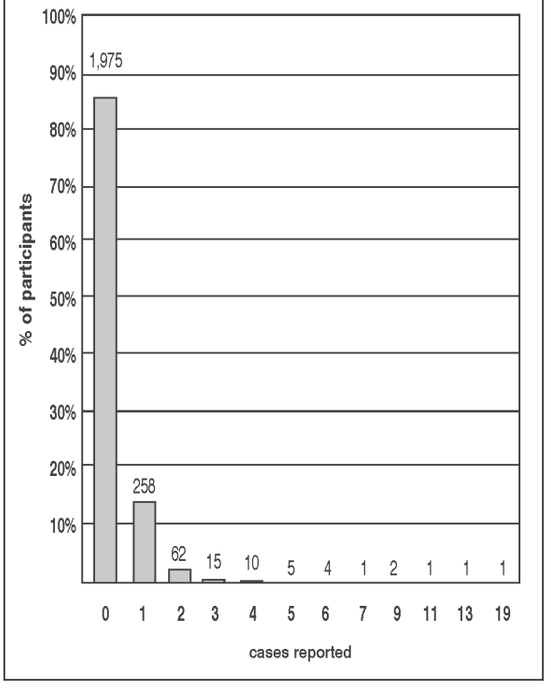


TABLE 2

Criteria considered for inclusion of studies

Rarity	Disorders of such low incidence or prevalence that national ascertainment of cases is needed (less than 1,000 cases a year).
Public health importance	Clearly addressing a public or paediatric health issue.
Scientific importance	Demonstrated scientific interest and importance.
Uniqueness	Proposal must demonstrate a clear need for data on a condition or disorder for which there is only limited information and for which surveillance is the most appropriate means of collecting the data.
Quality of proposal	Proposal must state clear and achievable objectives, practicability, patient confidentiality, adequate resources, clear questionnaire and method of evaluation.
Workload of paediatricians	Steering Committee must be convinced that reporting will not make excessive additional demands on the workload of paediatricians.
Priority will be given to diseases that are not currently notifiable or, if notifiable, have sufficient indication of under-notification. Investigators are expected to demonstrate that potential funding is available.	

reporting form. Once collected, results are forwarded to the investigator for data analysis.

Results of the 2003 one-time survey question on lap-belt syndrome are found on page 49.

Investigators' corner

The CPSP can offer investigators the use of a timely, active surveillance system to increase awareness of childhood disorders that are high in disability, morbidity, mortality and economic costs to society, despite their low frequency. The CPSP provides an innovative means of identifying and obtaining data on low-frequency diseases and conditions from approximately 2,335 participants. The program is committed to a case ascertainment rate of over 90% and boasts a high response rate of 96% on detailed reports (Table 1), due to follow-up reminders to participants who have not responded. 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 once they have reviewed

TABLE 3

Format for submission

Proposals for new studies should include:

- name of principal author
- brief abstract of proposal
- proposed starting date
- proposed duration
- question(s) to be addressed by study
- statement of justification, including how the information could be used
- case definition
- expected number of cases
- availability of ethical approval (state source of approval)
- funding arrangements
- identification of projected date for completion of analysis and submission for publication

the *Criteria considered for inclusion of studies* (Table 2) and the *Format for submission* (Table 3). The Steering Committee reviews submissions at its spring and fall meetings, giving preference to studies that have either strong public health importance or could not be undertaken any other way. Studies must receive ethical approval and have funding in place before final acceptance to the program.

The CPSP is pleased to see established faculty members mentoring young researchers with their study proposals.

As previously mentioned in the *Overview* section, the CPSP is available to investigators as a cost-effective tool to survey participants on a one-time basis in order to identify the prevalence of a problem or to answer a specific question.

Studies timeline

TABLE 4

CPSP studies timeline (by end date)

Studies	Start date	End date	Total confirmed cases to December 31, 2003
Group B strep	January 1996	December 1996	178
Neural tube defects	January 1997	December 1998	107
Creutzfeldt-Jakob disease	January 1997	June 1999	1
Hemorrhagic disease of the newborn	January 1997	December 2000	6
Subacute sclerosing panencephalitis	January 1997	December 2000	3
Cerebral edema in diabetic ketoacidosis	July 1999	June 2001	23
Progressive intellectual and neurological deterioration	July 1999	June 2001	59
Anaphylaxis	January 2000	June 2001	732
Hemolytic uremic syndrome	April 2000	March 2002	140
Smith-Lemli-Opitz syndrome	January 2000	December 2002	35
Hepatitis C virus infection	February 2001	January 2003	58
Neonatal liver failure/perinatal hemochromatosis	February 2001	January 2003	10
Necrotizing fasciitis	September 2001	August 2003	37
Neonatal herpes simplex virus infection	October 2000	September 2003	58
Neonatal hyperbilirubinemia – severe	July 2002	June 2004	203
Vitamin D deficiency rickets	July 2002	June 2004	69
CHARGE association/syndrome	September 2001	August 2004	90
Acute flaccid paralysis	January 1996	December 2004	370
Congenital rubella syndrome	January 1996	December 2004	9
Prader-Willi syndrome	January 2003	December 2004	31
Osteogenesis imperfecta	January 2004	December 2004	N/A
Early-onset eating disorders	March 2003	February 2005	63
Lap-belt syndrome	September 2003	August 2005	3
Adverse drug reactions – serious and life-threatening	January 2004	December 2005	N/A
Severe combined immunodeficiency	April 2004	March 2006	N/A
Acquired demyelinating syndromes of the central nervous system	April 2004	March 2007	N/A
Acute rheumatic fever	April 2004	March 2007	N/A

Program Evaluation

Report from the Chair of the Expert Advisory Group

Preamble

The CPSP evaluation process began early in 2003 with the creation of an Expert Advisory Group (EAG). Individual members were selected based on their experience and expertise in the fields of public health, paediatrics, epidemiology, surveillance and administration. The CPSP Evaluation Working Group held a preparatory meeting in May with the chair and circulated extensive background materials to all EAG members prior to a face-to-face meeting in September, followed by in-camera deliberations. The chair of the EAG presented the following final report to the CPSP Steering Committee in November 2003.

Members of the Expert Advisory Group

Dr. Robert McMurtry (Chair), Former Dean of Medicine, The University of Western Ontario; Former Assistant Deputy Minister of Health

Dr. Margaret Berry, Neonatologist, The Montreal Children's Hospital

Dr. Jeffrey Davis, Chief Medical Officer, Wisconsin Division of Public Health

Dr. Philippe Duclos, Project Leader, Immunization Safety, World Health Organization

Dr. Monika Naus, Epidemiologist, BC Centre for Disease Control; Chair, National Advisory Committee on Immunization

Overall comments

The EAG was unanimous in their opinion that the CPSP represents excellent value for money. The achievement in this respect was seen as excellent and unsurpassed by any comparable program known to the EAG. The CPSP was seen as representing an important collaborative tool for surveillance, research and policy development. In this role, it was perceived as unique in Canada. In other words, it provides an important activity that would disappear in its absence, unless a much larger investment is made to replace it.

The core activity of surveillance of low-frequency, high-impact conditions affecting children has created a network that reaches into all parts of Canada. This not only generates crucial information of these conditions ("they are on target"), but it has also established a mechanism to provide important public health information quickly and inexpensively on a national basis. Examples include the work on hemorrhagic disease of the newborn, confirming the Canadian recommendation of vitamin K as the gold standard for prevention, and on baby walker injuries, confirming the need for a commercial product safety ban on these devices.

The EAG was impressed by the survey of clinicians (paediatricians) that affirmed a change in practice pattern by some, and a high degree of recognition by all. The publications generated by the program also received accolades. The CPSP is encouraged to increase its reach to include nurse practitioners and northern communities and territories.

Finally, the EAG underlined the importance of providing more evidence of impact on public health policy and clinical practice. Annual evaluation of the effectiveness of the Steering Committee was also recommended.

Program objectives

The CPSP has done well in regards to its current objectives. It has initiated programs of national scientific significance and developed an effective surveillance system to monitor the health of Canadian children in relationship to low-frequency, high-impact conditions. Nonetheless, there may be an advantage to rewording the program objectives to reflect emerging priorities and new realities (e.g., changes in federal leadership, positive changes in federal/provincial/territorial relations).

Some specific wording for the program objectives was suggested as follows:

- to identify important disease conditions for surveillance to support paediatricians and public health officials in their role of contributing to the health and well-being of Canadian children;
- to ensure a strong infrastructure and maintain and improve a national and collaborative population-based surveillance system to monitor health in Canadian children;
- to facilitate research into low-frequency, high-impact childhood disorders for the advancement of knowledge, the enhancement of understanding and the improvement of treatment, prevention and health-care planning.

The EAG commended the CPSP on performing its core function so well and emphasized that important additional roles, such as responding to public health emergencies and international collaboration, may require additional resources.

Evaluation objectives

The evaluation process was seen as exemplary, and the EAG was impressed with the surveys of the four stakeholder groups and the CDC (Centers for Disease Control and Prevention) framework. The provided integrated background material was well done and contained both quantitative and qualitative information of value. The logic frameworks provided an interesting context. However, the program goals were not seen as serving CPSP well and could be deleted without ill consequence.

The case in support of the excellent value for money represented by the CPSP might be strengthened, especially in view of the new federal fiscal reality that will likely be similar to the Program Review of 1994-95. The EAG is convinced that the case can be made and, furthermore, an effort to duplicate the essential work of the program by another means would be considerably more expensive.

Strategic issues and conclusion

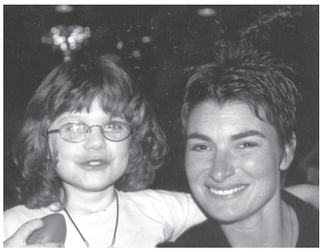
The events of 2003 have been characterized by large-scale change and high impacts. All provinces east of Alberta held elections this year with new governments being elected in Ontario, Quebec, and Newfoundland and Labrador. Most observers feel that together with the change in federal leadership, a more collaborative approach at federal/provincial/territorial forums can be anticipated. In addition, a significantly negative economic impact was felt from SARS and the case of one animal with BSE (bovine spongiform encephalopathy). Both were low-frequency, high-impact events and, accordingly, both of these latter developments underscore the importance of public health and the crucial importance of surveillance.

In the reviewers' opinion, the asset that the CPSP represents is relevant to these realities. It is a national program and an important mechanism for surveillance of human health as observed in the health and well-being of one of the most vulnerable populations in Canada, our children.

CPSP Principal Investigators

Surveillance studies in 2003

Dr. Paul Varughese
Acute flaccid paralysis
and
Congenital rubella syndrome



Dr. Kim Blake
CHARGE association/syndrome

Dr. Leora Pinhas
Early-onset eating disorders

Dr. Claude Cyr
Lap-belt syndrome

Dr. H. Dele Davies
Necrotizing fasciitis

Dr. Tom Wong
Neonatal herpes simplex virus infection

Drs. Michael Sgro and Vibhuti Shah
Neonatal hyperbilirubinemia – severe

Dr. Glenn Berall
Prader-Willi syndrome

Dr. Leanne Ward
Vitamin D deficiency rickets

New studies in 2004

Dr. Brenda Banwell
Acquired demyelinating syndromes of the central nervous system

Dr. Christina Templeton
Acute rheumatic fever

Dr. Bruce Carleton
Adverse drug reactions – serious and life-threatening

Dr. Leanne Ward
Osteogenesis imperfecta

Dr. Louise Pelletier
Severe combined immunodeficiency

Surveillance Studies in 2003

Acute flaccid paralysis

(January 1996 to December 2004)

Highlights

- No wild polio cases have been isolated in Canada since 1988.
- The number of AFP cases reported for 2003 and 2002 was lower than in previous years; however, duplicate reporting remains high.
- Guillain-Barré syndrome accounts for at least 74% of confirmed AFP cases.
- Polio viral stool cultures are still essential.

Background

The elimination of indigenous wild poliovirus transmission in Canada, and the rest of the American region, was certified in September 1994. However, until global polio eradication is attained, there remains an ongoing risk of wild poliovirus importation from polio-endemic regions to Canada. Consequently, active surveillance of acute flaccid paralysis (AFP) in children less than 15 years old is used to monitor potential cases of paralytic poliomyelitis. Based on World Health Organization (WHO) criteria for AFP surveillance (Table 5), the estimated minimum number of cases in Canada is 58 per year. AFP surveillance in Canada was initiated in 1991 through the IMPACT (Immunization Monitoring Program ACTive) network of paediatric tertiary-care centres and, since 1996, has been implemented through the CPSP. This report presents the results of AFP surveillance in 2003 and compares them to those from previous years.

Objective

The objective of AFP surveillance is to identify AFP cases (including Guillain-Barré syndrome [GBS]) in children less than 15 years of age to rule out paralytic poliomyelitis and thereby monitor the polio-free status of Canada.

TABLE 5

World Health Organization quality assurance criteria for acute flaccid paralysis surveillance

1) one case per 100,000 in a population less than 15 years of age
2) adequate polio virus stool culture in 80% of cases

Case definition

Acute onset of focal weakness or paralysis characterized as flaccid (reduced tone) without other obvious cause (e.g., trauma) in children less than 15 years of age. Transient weakness (e.g., post-ictal weakness) should not be reported.

Results and discussion

In 2003, the CPSP received 84 initial AFP reports, of which 42 (50%) were confirmed cases. The other half were split between 35 duplicate reports and seven cases that did not meet the AFP surveillance case definition or have adequate information. Forty-two confirmed cases represents a rate of 0.72% per 100,000 which is below the minimum estimated background rate of one case per 100,000 children less than 15 years of age, or 58 cases. With the anticipated 'late reports' for the current year, the final number is likely to be slightly higher, but below the WHO targeted rate. Cases ranged in age from two months to 15 years (median 5.9 years, mean 7.3 years) in 2003. Table 6 shows the age distribution of AFP cases reported in 2003 compared with cases reported from 1996 to 2002. Overall, the age distribution is similar throughout the reporting period. More than two-thirds (69%) of cases were males.

Although most Canadian children today are vaccinated against polio, only 29 of the 42 cases (69%) had documentation of routine childhood immunization, and all had received age-appropriate polio immunization. For the remaining 13 cases, no polio vaccine-specific information was available on the detailed case report forms.

Virological investigation for polio or other enteroviruses

A total of 20 cases (48%) had stool examination; virology was not done or the status was unknown for 22 cases (52%). However, adequate stool investigation for the isolation of poliovirus or non-polio enteroviruses (i.e., stool specimen collected within two weeks of the onset of paralysis) was reported for only 17 of 42 cases (40%). For three additional cases, although stool specimens were collected, it was after two weeks of onset of paralysis. None were positive for polioviruses; one was characterized as 'adenovirus' and another as 'echovirus'. None of the nine throat and/or 23 cerebrospinal fluid specimens collected for viral isolation was positive for poliovirus. For 17 of the 42 cases (40%), stool specimens were also tested for *Campylobacter* organisms; but all were negative.

Neurological investigations consisted of at least one or more of the following:

CSF abnormalities (protein, glucose, WBC, neutrophils, lymphocytes, and RBC), nerve conduction studies, electromyography, MRI or CT scan; abnormal findings compatible with the neurological diagnosis were reported for one or more of the tests done. Twenty-six (72%) of the 36 CSF specimens indicated some abnormal findings. MRI or CT scanning was done for 37 cases (88%); for MRI, 14/21 or 67% showed some abnormality. Electromyography and/or nerve conduction studies were done for 33 cases; 26 (79%) of which had abnormal findings. Guillain-Barré syndrome was the final neurological diagnosis in 31 cases or 73.8% (one was a GBS Miller-Fisher variant) and transverse myelitis in four (9.6%) (Table7). The remaining seven diagnoses included acute disseminated encephalomyelitis (3), lumbosacral plexineuropathy (1), progressive ischemic myelopathy (1), encephalomyelitis radiculitis (1), and post-infectious transverse myelitis (1).

Forty-one of the 42 required hospitalization for periods ranging from one to 105 days (mean of 17 days); seven cases were hospitalised for 30 days or longer. Of the 42 cases, two (4.8%) recovered fully, 29 (69%) recovered partially with residual weakness, and 11 (26. 2%) had an unknown recovery status at 60 days after the onset of paralysis.

None of the clinical specimens tested, i.e., stool, nasopharyngeal or cerebrospinal fluids, were positive for poliovirus infection.

TABLE 6

Age distribution of AFP cases reported to the CPSP, 1996-2003

Age group (years)	Number of cases (%)							
	1996	1997	1998	1999	2000	2001	2002*	2003
0 – 1	2 (6.7)	—	2 (4.6)	3 (4.9)	2 (3.3)	8 (14.8)	10 (23.3)	5 (11.9)
2 – 5	11 (36.7)	13 (37.1)	15 (34.1)	18 (29.5)	24 (39.3)	18 (33.3)	16 (37.2)	17 (40.5)
6 – 10	9 (30.0)	12 (34.3)	18 (40.9)	23 (37.7)	22 (36.1)	14 (25.9)	11 (25.6)	7 (16.7)
11 – <15	8 (26.6)	10 (28.6)	9 (20.4)	17 (27.9)	13 (21.3)	14 (25.9)	6 (13.9)	13 (30.9)
Total	**30 (100)**	**35 (100)**	**44 (100)**	**61 (100)**	**61 (100)**	**54 (100)**	**43 (100)**	**42 (100)**

* Includes four delayed reports not included in the *CPSP 2002 Results*.

TABLE 7

Neurological diagnosis of AFP cases reported to the CPSP, 1996-2003

Final Diagnosis	Number of cases (%)							
	1996	1997	1998	1999	2000	2001	2002*	2003
Polio	0	0	0	0	0	0	0	0
Guillain-Barré syndrome	21 (70.0)	29 (82.8)	34 (77.3)	50 (82.0)	49 (80.3)	42 (77.7)	33 (76.7)	31 (73.8)
Transverse myelitis	6 (20.0)	2 (5.7)	6 (13.6)	7 (11.5)	4 (6.6)	8 (14.8)	7 (16.3)	4 (9.6)
Encephalitis/encephalomyelitis/encephalopathy	1 (3.3)	1 (2.9)	1 (2.3)	—	—	—	—	3 (7.1)
Myelopathy	—	1 (2.9)	—	—	—	—	—	—
Radiculopathy/radiculoneuritis	1 (3.3)	1 (2.9)	—	—	—	1 (1.9)	1 (2.3)	—
Plexitis/lumbosacral plexitis	—	—	—	2 (3.2)	—	—	—	1 (2.4)
Brachial neuritis	—	—	—	1 (1.6)	—	—	—	—
Rhombomyelitis	—	—	—	1 (1.6)	—	—	—	—
Other	—	—	—	—	8 (13.1)	3 (5.6)	2 (4.7)	3 (7.1)
Not specified/undetermined diagnosis or etiology	1 (3.3)	1 (2.9)	3 (6.8)	—	—	—	—	—
Total	**30 (100)**	**35 (100)**	**44 (100)**	**61 (100)**	**61 (100)**	**54 (100)**	**43 (100)**	**42 (100)**

* Includes four delayed reports not included in the *CPSP 2002 Results*.

Conclusions

The 42 AFP cases identified to date for 2003 are below the expected rate in Canada, according to the World Health Organization criteria. For the corresponding period for 2002, a total of 39 cases were initially reported, although the final number has since increased to 43 with the inclusion of four additional 'late' reports.

The decline in the number of AFP cases documented by the CPSP over the past three years might be due to under-reporting of cases or epidemiological peculiarities: both need to be further investigated. However, the high number of duplicate case reports, and the continued involvement of IMPACT, may in fact indicate this to be a true reflection of the changing trend.

It is still encouraging to note that the AFP reporting rate has improved since the introduction of paediatrician-based reporting through the CPSP from 0.5 per 100,000 children less than 15 years of age in 1996 (30 cases) to 1.04 per 100,000 in 2000 (61 cases). For unknown reasons, the rate has decreased since then. Undoubtedly, the expansion of AFP surveillance to the CPSP has improved the completeness of surveillance by ensuring that AFP cases seen at non-tertiary hospitals are reported in addition to those cases admitted to paediatric tertiary care hospitals that are reported through IMPACT.

A major area in which the AFP surveillance could be improved is the performance of polio-specific investigations and timely reporting of results. The proportion of cases where polio-specific laboratory investigations were reported remained low in 2003, with only 48% of cases having had an adequate stool investigation during this period. This compares with the 33% to 51% reported for the period 1996-2002. These rates of adequate stool investigation remain significantly lower than the WHO target of 80%. While neurological investigations provide supporting evidence for the

final diagnosis in the majority of reported AFP cases, polio-specific laboratory investigations remain vital for the evaluation of all cases, including those in which poliomyelitis is not being considered as a possible diagnosis. Negative results of appropriate polio-specific investigations are as important as a positive result would be in AFP case evaluations. The single most important laboratory investigation, recommended by the National Working Group on Polio Eradication, to confirm or to rule out a diagnosis of paralytic poliomyelitis, is a stool specimen collected within two weeks of onset of paralysis for isolation of wild or vaccine strain poliovirus. Specimens may be collected up to six weeks after the onset of paralysis, although after two weeks, the sensitivity of virus isolation decreases. The examination of paired serum samples for evidence of a fourfold or greater rise in poliovirus antibody titre in paired sera and/or the presence of poliovirus-specific IgM antibody in a single serological specimen further enhance the evaluation of cases.

Principal investigator

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CHARGE association/ syndrome

(September 2001 to August 2004)

Highlights

- Between 2001 and 2003, eight to ten patients per year were identified with CHARGE A/S.
- Significant causes of morbidity and presumed mortality were severe feeding difficulties (often requiring surgical interventions), cardiovascular and CNS anomalies.
- Preliminary cohort data (13 individuals) suggest evidence of autism spectrum disorder in the majority.

Background

CHARGE association/syndrome (CHARGE A/S) is a constellation of a number of congenital anomalies that was first given the acronym CHARGE (Coloboma, Heart Defect, Choanal Atresia, Retarded Growth and Development, Genital Hypoplasia, Ear Anomalies/Deafness) in 1981.

Over the past 15 years, the specificity of this pattern of malformations has reached the level that many clinicians now consider it to be a discrete recognizable syndrome (Graham JM. *Am J Med Gen* 2001;99:120-3). With increasing expertise, it became clear that the criteria originally proposed needed further refinement. The revised consensus diagnostic criteria by Blake et al. incorporated both major and minor features for CHARGE A/S and have been documented to enhance clinical diagnosis and facilitate research efforts. These criteria consist of four major characteristics: coloboma, choanal atresia, characteristic ear anomalies, cranial nerve dysfunction (facial palsy, vestibular dysfunction, and swallowing difficulties) and seven minor criteria: heart defect, orofacial cleft, genital hypoplasia,

growth deficiency, developmental delay, tracheoesophageal fistula and a distinctive facial appearance. The diagnosis is firmly established when all four major or three major and three minor criteria are present. Some of the criteria are difficult to detect in infants, and as the major characteristics are rare in other conditions, the CHARGE A/S diagnosis needs to be considered in any individual who has one or two major criteria and several minor characteristics. To define CHARGE A/S in these individuals, a cranial CT or MRI scan may show hypoplasia of the semicircular canals and/or cochlea and/or choanal atresia or stenosis. High resolution chromosome studies, fluorescence *in situ* hybridization (FISH) for 22q11 deletion and the subtelomeric deletion FISH testing help to rule out any chromosomal abnormalities accounting for the multiple congenital anomalies. An increase in paternal age of CHARGE A/S children has been recognized as a risk factor and needs to be confirmed.

The purpose of this study is to determine the incidence and prevalence of CHARGE A/S in Canada, as the true incidence is unknown. As CHARGE A/S presents with a wide spectrum of clinical severity, mildly affected patients may also be diagnosed and can be followed prospectively. The review article, entitled "CHARGE association: An update and review for the primary paediatrician" (*Clin Pediatr* 1998;37:159-74), summarizes current understanding of the management of this complex and chronic multiple congenital anomaly, giving physicians a guide to the management of CHARGE A/S.

Objectives

1) To determine the incidence and prevalence of CHARGE A/S in Canada by ascertaining all identified cases of CHARGE A/S (old and new).
2) To obtain demographic and medical information on patients with CHARGE A/S, and assemble a database to answer research questions.
3) To follow developmentally and behaviourally an identified group of CHARGE A/S infants who have been diagnosed at an early age and have obtained early intervention services. Will early recognition and treatment of these infants improve their clinical and behavioural well being?

Case definitions

Infant/child/adult with four major criteria or three major and three minor criteria.

- Major inclusion criteria: coloboma, choanal atresia, characteristic ear abnormalities, cranial nerve dysfunction.
- Minor inclusion criteria: genital hypoplasia, developmental delay, cardiovascular malformations, growth deficiencies, orofacial cleft, tracheoesophageal (TE) fistula, characteristic face.

Exclusion criteria

Exclude other conditions such as velocardiofacial syndrome and DiGeorge Sequence using FISH test (fluorescent *in situ* hybridization) to exclude 22q11 deletion.

Results

Incidence

In 28 months of surveillance, 90 confirmed individuals, 48 males and 42 females, were reported with CHARGE A/S. Forty-three percent of the families agreed to be contacted for further follow-up studies. Table 8 not only provides current and cumulative data on case reports, but also demonstrates the high rate of duplicate reporting of CHARGE A/S cases.

Calculations of the estimated regional incidence for CHARGE A/S continued to demonstrate a provincial variation. The highest incidence was from provinces where the awareness of CHARGE A/S is particularly high, the Atlantic Provinces, Manitoba and Saskatchewan; averaging 1:9,300 live births with

TABLE 8

CHARGE association/syndrome cases

	Reported	Confirmed	Duplicates	Discards	Pending
2003 (12 mths)	37	12	12	11	2
2001-03 (28 mths)	174	90*	51	31	2

* In the 28 months of surveillance, the Canadian estimated incidence of CHARGE A/S is 1:26,700.

CHARGE A/S. The global Canadian incidence is estimated at 1:23,000 live births.

Figure 4 demonstrates that a higher proportion of individuals with CHARGE A/S were identified within the younger population. There is still a paucity of adolescents being identified through the surveillance.

Morbidity and mortality

Feeding problems were present in 83% (75/90) of cases; gastrostomy and/or jejunostomy tubes were required in 77% (61/79). Among deceased individuals, gastroesophageal reflux (GER) was present in 100% (6/6) of individuals older than one month at the time of death. See Table 9 for further mortality details. Major cardiovascular anomalies were present in 70% of the deceased population compared to 35% of the surviving cohort, with conotruncal anomalies and atrioventricular septal defects (AVSD) being significantly more common in the deceased compared to the surviving CHARGE A/S population.

Structural anomalies of the central nervous system, as diagnosed by computed tomography or magnetic resonance imaging, were more prevalent in the deceased compared to the surviving population (70% vs. 51%).

Behaviour and development

Results of standardized parent questionnaires on development/behaviour and a structured telephone interview are reported on the first 13 individuals (eight males, five females). This population has relatively low adaptive behaviour skills, motor impairment being particularly significant (Table 10).

TABLE 9

Deceased patients with CHARGE association/syndrome

	Age at time of death	Sex	Choanal atresia	Feeding difficulties	Cardiovascular defects	Other anomalies
1	<1 week	F	BPCA	GER, G-tube	TOF	H, A, N
2	7 weeks	M	—	GER, G-tube	PDA, ASD, TR	R, H, N
3	6.5 months	F	BPCA	GER, G-tube	PDA, ASD	—
4	8 months	F	—	GER, vomiting	TOF, AVSD	TEF, R
5	1 week	F	BPCA	—	PDA, abn TV & PV, RVH	F
6	4.5 months	F	CS	GER, aspiration, G-tube	AVSD, absent PV	F
7	2.5 months	M	CS	GER, partial nasal obstruction	—	CP, R, H, N
8	1 week	F	BPCA	TEF, G-tube	PS	TEF, R
9	9 years	M	CA	GER, G-tube	DORV, subaortic stenosis	CL, CP, F, R, N, T
10	2 weeks	M	—	—	DORV, AVSD, PS	F, N

A: abdominal defects; A(V)SD: atria (ventricular) septal defect; BPCA: bilateral posterior choanal atresia; CL/P: cleft lip/palate; CS(A): choanal stenosis (atresia); DORV: double outlet right ventricle; F: characteristic facial features; GER: gastroesophageal reflux; H: hand anomalies; N: neck/shoulder anomalies; PDA: patent ductus arteriosus; PV(S): pulmonary valve (stenosis); R: renal anomalies; RVH: right ventricular hypertrophy; T: teeth anomalies; TEF: tracheoesophageal fistula; TOF: tetralogy of Fallot; TV(R): tricuspid valve (regurgitation).

TABLE 10

Vineland Adaptive Behavior Scales standard scores of CHARGE A/S individuals

Case #	Age at interview (yr-mo)	Communication	Daily living skills	Socialization	Motor skills*	Adaptive behaviour composite
1	3-2	60	56	59	50	52
2	3-5	82	62	89	56	67
3	4-4	79	80	84	51	68
4	5-7	48	50	56	45	46
5	4-6	68	56	80	50	58
6	4-4	53	53	66	56	52
7	5-4	57	59	67	58	55
8	5-10	43	38	52	—	41
9	9-1	26	43	50	n/a	37
10	12-1	<20	<20	<20	n/a	<20
11	17-3	111	88	46	n/a	56
12	23-7	†	†	†	n/a	†
13‡	24-2	16-6	18-0	12-8	n/a	15-8
Mean§	**6-10**	**59.8**	**55.4**	**61.2**	**52.3**	**50-5**
SD	(4-4)	(27.3)	(19.8)	(20.9)	(4.6)	(14.7)

* Motor skills administered only for children less than 8 years of age.
† Missing data
‡ Above age range of standardization sample; age-equivalent scores are shown.
§ Expected mean = 100, SD = 15

TABLE 11

Social-Communication Questionnaire (SCQ) scores* and clinical judgments of evidence for autism spectrum disorder (ASD) for CHARGE A/S individuals

Case #	Verbal†	SCQ score‡	Evidence of ASD
1	no	10	some, but <4 years
2	yes	10	some, but <4 years
3	yes	6	little
4	no	8	some
5	yes	10	moderate
6	no	12	some
7	no	8	some
8	no	16	moderate
9	no	13	strong
10§	no	23	n/a
11	yes	12	moderate
12	no	29	moderate
13	yes	14	strong

* Berument et al., 1999
† Refers to whether child is able to talk using short phrases and sentences.
‡ In verbal individuals, scores of 15 and higher are associated with an increased probability of autism, whereas in non-verbal individuals a lower SCQ cutoff may be accepted.
§ Severe-profound hearing/visual impairments, mental retardation – unable to assess for ASD.

FIGURE 4

Age distribution of CHARGE association/syndrome in Canada

28 months of surveillance

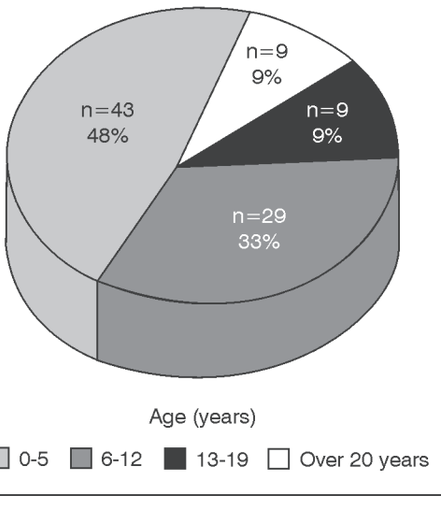

TABLE 12

Confirmed cases of CHARGE association/syndrome born between 1995-2003

Reporting province	1995	1996	1997	1998	1999	2000	2001	2002	2003
Alberta	1	0	0	0	0	0	0	0	0
British Columbia	1	0	1	0	3	1	3	1	0
Manitoba	0	0	0	2	2	0	0	1	1
Maritimes*	1	0	0	5	1	2	0	0	0
Newfoundland/Labrador	0	0	0	0	0	0	0	2	0
Ontario	0	1	4	5	1	2	3	3	4
Quebec	0	1	3	2	1	2	1	3	0
Saskatchewan	0	0	0	0	1	0	2	0	3
Total cases per year	**3**	**2**	**8**	**14**	**9**	**7**	**9**	**10**	**8**

* Maritime provinces include New Brunswick, Nova Scotia and Prince Edward Island.

The majority showed evidence of autism spectrum disorder (ASD) although it is important to recognize the inherent challenges in diagnosing autism in individuals with sensory impairments.

Figures in Table 12 represent confirmed cases of CHARGE A/S born within the last nine years; not included in these figures are cases that are still pending confirmation of diagnosis, or the many duplicate reports that were received. If clinicians are unsure if their case has been reported, they can use this as a guide. Duplication is always better than to allow the chance of missing a report. Nine or ten cases per year yields an incidence of 1:30,000.

Conclusions

Ninety individuals represent the largest CHARGE A/S cohort in the literature. In several provinces where the awareness of CHARGE A/S is particularly well developed, an estimated incidence approaches 1:9,000 live births. As this study has identified a high prevalence of feeding difficulties that contribute significantly to the mortality and morbidity issues for CHARGE A/S individuals, investigation and intervention for GER and associated feeding difficulties are recommended. It will be important to compare both rates and presentation of autism spectrum disorder in individuals with CHARGE A/S to those seen in other congenital anomaly syndromes, as well as in persons with sensory impairments.

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Congenital rubella syndrome

(January 1996 to December 2004)

Highlights

- There were no new CRS cases in 2003.
- From 1996 to 2003, zero to two newborns with CRS per year were identified through the surveillance systems in Canada (0 to 0.5 per 100,000 births).
- Canada's very low incidence of rubella and CRS is a reflection of the impact of rubella elimination strategies.
- This study reinforces the need for all children to receive their rubella vaccine at the recommended ages.
- Standing orders for vaccination of all rubella susceptible women in the immediate postpartum period are essential.

Background

In Canada, rubella immunization programs were introduced in the 1970s. However, the program strategies varied; some provinces initially opted for selective immunization of pre-adolescent females and others opted for immunization of all infants. By 1983, all provinces and territories across Canada had implemented routine measles-mumps-rubella combined vaccine (MMR) at 12 months. During 1996 and 1997, all provinces and territories introduced a routine second dose MMR or measles-rubella combined vaccine (MR) given at 18 months or four to six years. Some jurisdictions used MR vaccine for their second dose catch-up campaigns.

Since 1970 the incidence of rubella in Canada has declined markedly; fewer than 30 cases were reported annually in the past three years. During a national consensus conference in 1994, a goal of eliminating indigenous rubella infection during pregnancy by the year 2000 was established. In November 2001, a National Expert Working Group on Rubella recommended that all rubella infections be included for enhanced surveillance.

In Canada, passive reporting of congenital rubella syndrome (CRS) to the Notifiable Diseases Reporting System (NDRS) began in 1979. Active surveillance of CRS began in 1992 through a network of tertiary-care paediatric hospitals (now representing more than 90% of paediatric tertiary-care beds in Canada) participating in IMPACT (Immunization Monitoring Program ACTive) and through the CPSP since 1996.

Objectives

1) To estimate the incidence of congenital rubella syndrome.
2) To obtain detailed epidemiological data, including maternal histories, on reported cases of congenital rubella syndrome and infection.

Case definitions

Confirmed case

Live birth

Two clinically compatible manifestations (any combination from Table 13, columns A and B) with laboratory confirmation of infection:

- isolation of rubella virus from an appropriate clinical specimen;

or

- detection of rubella-specific IgM in the absence of recent immunization with rubella-containing vaccine;

or

- rubella-specific IgG persisting at elevated levels for longer than would be expected from passive transfer of maternal antibody, or in the absence of recent immunization.

Stillbirth

Two clinically compatible manifestations with isolation of rubella virus from an appropriate clinical specimen.

Note: The following cannot be classified as a CRS case:

- rubella antibody titre absent in the infant;

or

- rubella antibody titre absent in the mother;

or

- rubella antibody titre declining in the infant consistent with the normal decline after birth of passively transferred maternal antibody.

Congenital rubella infection
Confirmed case
A case with laboratory confirmation of infection but with no clinically compatible manifestations:

- isolation of rubella virus from an appropriate clinical specimen;

or

- detection of rubella-specific IgM in the absence of recent immunization with rubella-containing vaccine;

or

- persistence of rubella-specific IgG at elevated levels for longer than would be expected from passive transfer of maternal antibody, or in the absence of recent immunization.

Rubella in clinical illness
Confirmed case
Laboratory confirmation of infection in the absence of recent immunization with rubella containing vaccine:

- isolation of rubella virus from an appropriate clinical specimen;

or

- significant rise in serum rubella IgG antibody levels by any standard serological assay;

or

- positive serologic test for rubella-specific IgM;

or

- clinical illness* in a person who is epidemiologically linked to a laboratory confirmed case.

* *Clinical illness is characterized by fever and rash, and at least one of the following: arthralgia/arthritis, lymphadenopathy, conjunctivitis. Up to 50% of rubella infections are reported to be subclinical.*

Results and discussion

In 2003, there were two reports of the same 20-month-old adopted child from Asia, who had the following clinical conditions: congenital cataract, patent ductus arteriosus, and microcephaly. No information was available on the mother. This case did not meet the Canadian CRS case definition as the diagnosis was confirmed beyond infancy. The very low incidence of CRS and rubella infection suggests that Canada is getting closer to achieving the goal of eliminating indigenous rubella infection during pregnancy.

TABLE 13

Congenital rubella syndrome: clinically compatible manifestations

Column A	Column B
1. Cataracts or congenital glaucoma (either one or both count as one) 2. Congenital heart defect 3. Sensorineural hearing loss 4. Pigmentary retinopathy	1. Purpura 2. Hepatosplenomegaly 3. Microcephaly 4. Micro-ophthalmia 5. Mental retardation 6. Meningoencephalitis 7. Radiolucent bone disease 8. Developmental or late onset conditions, such as diabetes and progressive panencephalitis and any other conditions possibly caused by rubella virus

From January 1996 to December 2003, with active surveillance in place, nine new reports of newborns with CRS were reported in Canada (Table 14). Of those whose status was recorded, four were born to immigrant women, one to an aboriginal woman, and two to non-aboriginal women. These seven cases illustrate the need for documentation of previously

TABLE 14

Cases of CRS by year of birth reported to CPSP/IMPACT and NDRS from January 1996 to December 2003

Year of birth	Reported to NDRS only	Reported to CPSP only	Reported to both NDRS and CPSP	Total
1996	1	0	1	2
1997	0	0	1	1
1998	0	0	1	1
1999	0	0	1	1
2000	0	0	2	2
2001	0	0	0	0
2002*	0	1	1	2
2003*	0	0	0	0
Total	**1**	**1**	**7**	**9**

* Notifiable Diseases Reporting System data is provisional.

received rubella vaccination, of maternal immunity status by a reliable method, and postpartum rubella vaccine when indicated.

Conclusions and recommendations

The very low incidence of CRS and rubella infections suggest that Canada is getting closer to achieving the goal of eliminating indigenous rubella infection during pregnancy.

Health-care providers are requested to ensure that: (1) all patients receive their rubella vaccinations at the recommended age and (2) all women without documented proof of rubella immunization receive the vaccine. Special attention should be given to the immunization status of women from regions with poor vaccination coverage, including women in immigrant populations. Routine rubella antibody screening antenatally by a reliable method is central to the congenital rubella prevention strategy, and all women found to be susceptible should be vaccinated in the immediate postpartum period. Standing orders for vaccination of susceptible women before discharge from hospital are the most effective way to ensure that the opportunity is not missed.

The degree of under-diagnosis and under-reporting for congenital rubella infection (CRI), CRS with less severe manifestations and CRS with delayed-onset manifestations is unknown. Physicians are reminded that it is important to investigate all infants born to mothers who have confirmed or suspected rubella infection during pregnancy, even if the infants have no obvious abnormalities on examination. Prenatal rubella screening and postpartum vaccination will continue to be essential in the quest to eliminate rubella infection during pregnancy.

Principal investigator

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Early-onset eating disorders

(March 2003 to February 2005)

Highlights

- Eating disorders were identified in 63 children aged 5-12 years old.
- Female to male ratio is 5 to 1 in 5- to 12-year-olds compared to 10 to 1 in older age groups.
- The predominant clinical feature (100%) was food avoidance.
- A substantial weight loss was documented during these important growing years.

Background

Over the last 50 years, the prevalence of anorexia nervosa in children and young adolescents appears to have been increasing while the age of onset of eating disorders has become even younger. However, there is ongoing debate in the literature about how to apply the current diagnostic criteria for eating disorders to children and younger adolescents. What is known though is that significant medical and psychological complications arise from starvation, weight loss or lack of appropriate weight gain during childhood and adolescence, making this group of conditions important to recognize and treat appropriately.

This study will document the incidence of early-onset eating disorders (EOED) in Canadian children and provide descriptive data on the abnormal cognitions, behaviours and severity of weight loss and/or other medical sequelae. This data will allow for a better understanding and recognition of this condition in younger children where currently the diagnosis may be delayed or missed. It will also aid in resource allocation and ultimately promote the creation of developmentally appropriate management guidelines to provide improved outcomes.

Objectives

1) To describe the minimum estimated incidence of early-onset eating disorders in children and young adolescents aged five to 12 years in Canada.
2) To describe the range of medical and psychiatric clinical features at presentation.
3) To compare the clinical features in children and young adolescents with existing diagnostic criteria for eating disorders in older patients.
4) To describe current therapeutic interventions used in management.

Case definition

Any child from five to 12 years of age inclusively seen in the previous month, with newly diagnosed early-onset eating disorder where eating disorder is defined as:

- determined food avoidance

and

- weight loss or failure to gain weight during a period of expected growth, not due to any identifiable organic cause such as celiac disease.

Exclusion criteria

- Obese children in a supervised weight management program

Results

- Number of confirmed cases is 63
- Estimated incidence is 1.95/100,000
- Female to male ratio is 5:1 (52 girls and 11 boys)
- Mean weight loss is 6.8 kg (± 4.7)

TABLE 15

Ethnicity

Ethnicity	Frequency n (%)
Asian	4 (6%)
Caucasian	55 (89%)
Mixed race	3 (4%)
Unknown	1 (1%)
Total	**63 (100%)**

TABLE 16

Symptoms or signs present at time of presentation

Symptom	Yes	No	Don't know
Food avoidance	62 (100%)		1
Excessive exercise	36 (58%)	26	1
Self-induced vomiting	7 (11%)	55	1
Fear of weight gain or fat	48 (86%)	8	7
Perception that body is larger	39 (72%)	15	9
Preoccupation with weight	48 (84%)	9	6
Preoccupation with food	53 (88%)	8	2
Laxative use	0	63	0
Diuretic use	0	63	0
Somatic complaints	17 (27%)	45	1
Denial of severity of symptoms	33 (65%)	18	12
Smoking	0	62	1
Weight loss	51 (89%)	6	6

TABLE 17

Comorbid psychiatric diagnosis

Diagnosis	Yes	No	Don't know
Depression	7 (12%)	51	5
Obsessive compulsive disorder	4 (7%)	53	6
Anxiety	18 (32%)	38	7
Other psychiatric illness	8 (13%)	55	0
Psychopharmacologic medication used	10 (17%)	48	5
Family psychiatric history	13 (22%)	47	3
Change in social situation	24 (40%)	36	3

Conclusion

Paediatricians and psychiatrists are identifying children and younger adolescents with eating disorders. The female to male ratio for the diagnosis of an eating disorder in young children between the ages of five and 12 years is 5:1 compared to 10:1 in

TABLE 18

Physical effects of disorder

Physical symptom	Yes	No	Don't know
Hypothermia	8	51	4
Hypotension	11	51	1
Bradycardia	18	44	1
Admitted to hospital	30	33	0
Nasogastric tube used	5	44	14
Patient alive	60	0	3

TABLE 19

Health-care providers involved

Providers	Yes	No	Don't know
Paediatrician	61	1	1
Psychiatrist	34	19	10
Dietitian	49	7	7
Psychologist	34	17	12
Social worker	36	16	11
Nurse	6	57	
Child/youth worker	6	57	
Family physician	1	62	

the older adolescent and adult population. Boys are more likely to be affected in the younger age group. This is the first data set on Canadian children to support the work of three previously published European studies. Although the majority were Caucasian, Asian and mixed race children were also identified. The group's mean weight loss of 6.8 kg (± 4.7) is a substantial weight loss in children between the ages of five and 12 years who should be gaining weight during these important growing years. Almost half (48%) required an inpatient admission for treatment. The average weight loss was 6.82 kg (±4.8) and 6.75 kg (±4.2) in girls and boys, respectively. While the greatest weight loss was found in older children aged 11 to 12 years of age, girls in this age group lost an average of 8.15 kg (+4.9) with a range of one

TABLE 20

Percent of children with weight loss by age and sex

Sex	5 to 8 years old	9 to 10 years old	11 to 12 years old
Boys	0/1 (0%)	3/3 (100%)	7/7 (100%)
Girls	2/3 (67%)	8/11 (73%)	29/38 (76%)

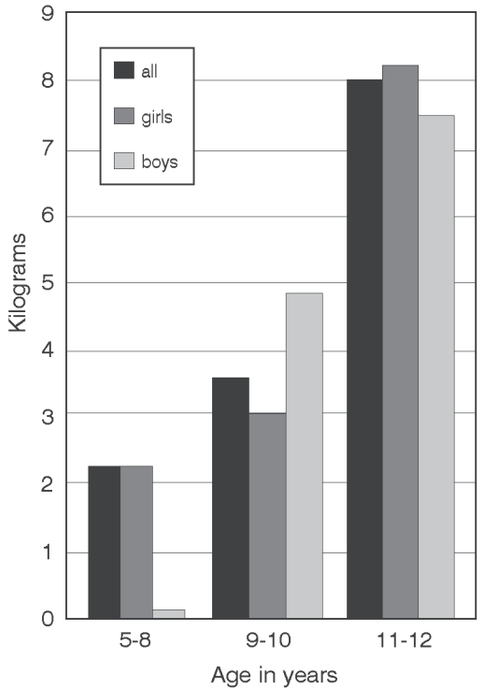
to 21 kg. This was approximately 18% of their total body weight. Boys aged 11 and 12 years old lost an average of 7.57 kg (+4.6) with a range of two to 15 kg. This was approximately 17.5% of their total body weight. Ninety-four percent of cases were identified in children over the age of eight years old. In that group, all of the boys (10/10), but only 76% (37/49) of the girls, had lost weight as part of their symptomatology.

Food avoidance was a predominant clinical feature in all confirmed cases. Many children also displayed a preoccupation with food and weight, and a fear of gaining weight. However, like their older adolescent counterpart, over half of these children denied their symptoms. Finally, one of the criteria for diagnosing anorexia nervosa in females is amenorrhea. Children under 12 years old would not necessarily be at an age where menstruation would be expected and, therefore, the criteria of amenorrhea for three consecutive months may not be useful for the diagnosis of anorexia nervosa in children and younger adolescents. Consequently, the majority of the children did not meet the full criteria for anorexia nervosa or bulimia nervosa as outlined in the DSM-IV.

This data suggests that it may be difficult to apply these diagnostic criteria to the younger age group. There may also be problems in matching clinical populations to the existing classification systems that are based on adults with eating disorders. Children have limited ability for insight that would be required to endorse the full spectrum of symptoms. Furthermore, children and younger

FIGURE 5

Average weight loss in kilograms by age and sex



adolescents may present with other types of clinical eating disturbances that are different from the classic eating disorders of anorexia nervosa and bulimia nervosa with respect to core psychopathology. Nonetheless, the presenting symptoms are as medically and psychologically problematic. Interestingly, some children do endorse the symptoms of vomiting (11%) and fear of gaining weight (86%) that have been thought to be only observed or reported in adolescents. Therefore, since these symptoms are unexpectedly higher than was originally hypothesized, further analysis is required.

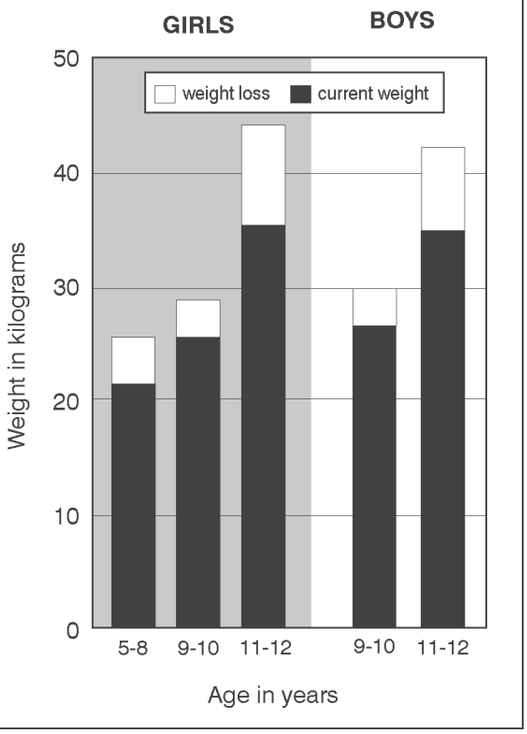
The majority of children did not have a history of a comorbid psychiatric diagnosis or a positive psychiatric family history. However, 40% had changes in their social situation. Bradycardia (28%) was the most common medical complication identified, and over 52% of the children were admitted to hospital.

This study is based on a similar data collection undertaken by the Australian Paediatric Surveillance Unit (APSU). After 21 months of surveillance, 45 cases of early-onset eating disorder in Australian children aged five to 13 years (inclusively) have been confirmed. Of the reported cases, 71% are female and 16% are younger than 11 years of age. A decrease in weight in the six months prior to diagnosis was observed in 89% of cases, with a median weight loss of 6 kg.

The profile of clinical features for these identified cases at the time of diagnosis is consistent with those being reported in Canada. However, abnormal

FIGURE 6

Weight loss compared to current weight in kilograms by age for girls and boys



medical findings are being reported in a higher proportion of Australian children with temperature less than 35.5°C reported in 40%, and bradycardia (minimum 36 beats/minute) reported in 53% of children. Concurrent depression is also being reported in a higher proportion of Australian children (40%).

Some international variation in management practices may be emerging, with 60% of the Australian sample receiving nasogastric (NG) feeding. This may be a consequence of a slight variation in reporting criteria during the first year of the APSU study which required only reporting children who were hospitalized. This might explain, in part, why there is a greater proportion of NG feeding reported by the APSU compared to the CPSP study. Surveillance through the APSU will continue until at least 2005.

International comparisons of the data from the EOED studies will enhance our knowledge of this global problem and will contribute to our understanding of early-onset eating disorders throughout the world.

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Lap-belt syndrome

(September 2003 to August 2005)

Highlights

- Seat belts were designed to save lives, but worn incorrectly they can cause important abdominal and lumbar spine injuries.
- Three cases of lap-belt syndrome were reported in the first four months, including one child with spinal cord injury.
- Media, public health and general interest in the study has been high.

Background

The use of seat belts has clearly reduced fatalities and severity of injuries in motor vehicle crashes. With the increasing use of seat belts over the last decades, a new association of injuries has emerged among adults and children involved in motor vehicle crashes. The 'seat-belt syndrome' was first described by Garrett and Braunstein in 1962 and refers to injuries to the intestinal viscera and to the lumbar spine associated with seat-belt restraints. Children are especially vulnerable to these injuries as their intra-abdominal organs are less protected by the thorax and pelvis, they have a lower centre of gravity and their iliac crests are less developed than those of adults allowing the belt to ride up over the abdomen. To date, there have been very few paediatric studies on the incidence of seat-belt syndrome. In fact, most current knowledge comes from case reports or studies done in limited regional areas. In these studies the number of cases was relatively low, ranging from ten to 50 cases over years.

Objectives

1) Obtain epidemiologic data on the incidence and pattern of injuries encountered in the seat-belt syndrome.
2) Identify at risk age groups.

3) Supply data that will help develop new strategies in order to adequately protect children in motor vehicles.
4) Promote education and awareness of this rare disease among health-care professionals.

Case definition

Any child up to and including 18 years of age restrained in a motor vehicle at the time of a crash, with either an abdominal injury, as determined by operation or CT-scan, or thoraco-lumbar spine injuries with or without spinal cord injuries.

Results and discussion

The study on lap-belt associated injuries started in September 2003. In the first four months of the study, Canadian paediatricians reported eight cases of lap-belt syndrome. Of these, three were confirmed, two were duplicates, one was a discard, and two had injuries probably related to their seat belts, but which did not meet the case definition (one child was in a booster seat and had a cervical spine fracture, while the other presented with a brachial plexus injury). Of the three confirmed cases, one presented with abdominal abrasions and a splenic contusion, another also had an abdominal abrasion and a jejunal infarct that needed small bowel resection, while the last presented with the full spectrum of seat-belt associated injuries, including an intestinal perforation that needed resection and an L2-L3 lumbar spine fracture with spinal cord injury.

Lap-belt syndrome is an important health problem in the Canadian paediatric population. It is associated with improper use of the lap belt only. Most encouragingly, even though cases have just started to be reported, interest from different media and public health organizations has been high. Issues pertaining to child safety in motor vehicles and the use of booster seats have been discussed not only with radio and newspaper journalists but also with the Ontario Ministry of Transport. The general public is already being informed and some provinces are taking legislative measures to ensure that children are adequately protected in motor vehicles. Clearly the most important issue of this study is to demonstrate the need to review restraints in motor vehicles. It is most satisfying to see that this objective is already being fulfilled.

Principal investigator

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Necrotizing fasciitis

(September 2001 to August 2003) Final report

Highlights

- Sixteen of the 26 (61%) GAS-related necrotizing fasciitis cases were varicella-associated.
- Implementation of universal varicella immunization would decrease NF incidence.
- Excruciating local pain out of proportion to skin presentation warrants urgent attention.
- Mortality was low but morbidity was high.
- All cases required surgery.

Background

In 1999, the Canadian Paediatric Society issued a statement on the state of knowledge and management of children and close contacts of persons with all-invasive group A ß-hemolytic streptococcal (GABHS) infections. In that statement, it was noted that there was no national data for necrotizing fasciitis (NF) in Canada. The current study started in September 2001 and concluded in August 2003 using the Canadian Paediatric Surveillance Program (CPSP) to establish actual national rates and epidemiology of NF.

Objectives

To define the epidemiology, management and outcome of NF in Canadian children.

Case definitions

NF is a deep-seated infection of the subcutaneous tissue that results in progressive destruction of fascia and fat. In general, NF is classified into two types.

1) Type I NF refers to mixed infections involving anaerobes (most commonly *Bacteroides* and *Peptostreptococcis spp*) and one or more facultative anaerobes, such as streptococci (non-GABHS), and members of the *Enterobacteriaceae* (e.g., *Escherichia coli*, *Enterobacter, Klebsiella, Proteus*).
2) Type II NF refers to that caused by invasive GABHS.

Results

Thirty-seven cases of NF were reported (26 group A streptococcus [GAS] related) across Canada with a mean age of 6.3 ± 4.9 years. Cases were reported from all areas of Canada west of the Maritimes. The annual incidence per million population peaked during the first year of life: age less than one year, 8 cases; 1-9 years, 3.4 cases; and 10-18 years, 1.1 cases, ($P<0.001$). Males under the age of one had the highest burden of disease with 12 cases per million population versus 3.2 cases per million for females ($P<0.0001$). In contrast, rates were similar between males and females for older age groups, (1-9 years: males, 3.2 cases, and females, 3.7 cases; 10-18 years: males, 1.0, and females, 1.25 cases, P=NS). Seventeen (46%) of all cases occurred within one month of the child developing varicella. Sixteen (61%) of the GAS related type II NF cases were varicella-associated versus one (9.1%) of the non-GAS type I related cases ($P<0.002$). Eight (21.6%) patients had an underlying chronic medical condition (2/26 type II related versus 6/11 type I related cases, $P<0.01$). For the 26 cases of type II related NF, the most common presenting symptoms were: localized pain (25, 96.2%), fever (10, 38.5%), vomiting (9, 34.6%) and chills (8, 30.8%). Localized pain was also reported uniformly in type I cases (11, 100%) along with chills (6, 54.5%) and fever (3, 27.1%). Eleven (42%) of the type II patients gave a history of having taken non-steroidal anti-inflammatory drugs (NSAIDS) within a week of admission. One type II patient had been in contact with another patient with invasive GAS disease. Two (18.2%) of the type I cases and none of the type II cases were nosocomial. Eighteen (48.6%) cases involved the lower extremities and/or groin area; nine (24.3%) the upper extremities (no type I cases involved upper extremities); nine (24.3%) the head, neck and chest areas; and two (5.4%) involved the abdomen (some patients had more than one site involved). All patients received at least one surgical procedure including fasciotomy/fasciectomy, debridement, amputation or skin graft. All type II

patients received clindamycin in combination with either penicillin (20 patients) or other beta-lactam antibiotic (6 patients). Overall, seven of the 37 patients (5/26 type II and 2/7 type I) were noted to have received IVIG, while another 14 (10/26, 38.5% type II and 4/11, 36.4% type I) patients received unspecified blood products. The mean length of stay was 16.8 ± 15.8 days. Two patients died (0/26 type II and 2/11, 18.2% type I).

Discussion

NF occurred throughout Canada, with the peak incidence among males younger than one year of age. The majority of type II NF cases were associated with recent varicella infection, whereas type I cases were more likely to be associated with underlying disease or be nosocomial. The lower extremities and groin areas were the most commonly involved parts of the body. Although outcomes with regard to mortality were excellent, there was substantial morbidity in the form of surgical procedures and prolonged hospital stay. This improved mortality may be related to one or more of the following: universal surgical intervention, early use of antibiotics and use of IVIG.

Conclusions

Surveillance through the CPSP has led to a better understanding of the epidemiology of paediatric NF in Canada. Pain out of keeping with clinical findings is an early symptom for clinicians. Type I and type II NF differ in terms of predisposing risk factors, relationship to varicella and possibly prognosis. This morbidity associated with type II NF supports the need for a universal varicella immunization program in Canada. Even though the largest at-risk population is under one year of age, "herd immunity" would be expected to reduce NF rates in this population.

Principal investigator

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Neonatal herpes simplex virus infection

(October 2000 to September 2003) Final report

Highlights

- Nine children died, resulting in a case fatality rate of 15.5%.
- Eight of 20 (40%) women were unaware of a history of HSV infection prior to delivery.
- The majority of typed cases (62.5%) were HSV-1, which has implications for vaccine and drug development.

Background

Herpes simplex virus (HSV) infections pose a public health concern especially since a high proportion of these infections are unrecognized. The most serious direct consequence of genital HSV infection is the perinatal transmission from mother to infant. With only limited data available, it is not possible to accurately determine the prevalence, incidence and trends of neonatal herpes infection in Canada. Data collection is essential to better understand the epidemiology of neonatal herpes and to monitor the trends in Canada. Canadian data on morbidity, mortality and on mother and infant risk determinants will allow comparison of neonatal herpes infection rates with other countries. The pre-vaccine baseline data that will be collected through this CPSP study before an HSV vaccine becomes available will be used to promote prevention and control program strategies, to further research, and to estimate the burden of illness in Canada.

Objectives

1) To estimate the incidence rate of neonatal herpes infections (HSV-1 and HSV-2) for the years 2000 to 2003 per 100,000 live births in Canada.
2) To determine the proportion of HSV-infected infants with localized and disseminated disease.

3) To identify risk determinants in mothers and their maternal HSV status prior to delivery.
4) To analyze trends of cases reported over a minimum period of three years, by age, sex and province.
5) To document the morbidity/mortality of neonatal infections for the years 2001 to 2006 through a cohort study of infants identified in each of the first three years of the neonatal herpes surveillance project.

Case definition

For the purpose of this study, the neonatal period was extended to 60 days of life so that late diagnoses were not missed, thereby optimizing the capacity to identify the maximum number of cases. All cases were laboratory-confirmed by at least one of the following tests:

1) Culture
2) HSV IgM
3) Polymerase chain reaction (PCR)

in an infant equal to or less than two months (60 days), born between October 1, 2000 and September 30, 2003, who demonstrates one of the following:

- localized infection involving the skin, eyes or mouth,
- disseminated infection:
  a) to central nervous system (CNS)
  b) to organs other than CNS.

Results and discussion

The status of all cases reported during the three-year study period was resolved as outlined below in Table 21.

A total of 58 cases of neonatal herpes simplex infection were confirmed (5.9 per 100,000 live births), nine of which were fatal. The majority of cases were reported from Central Canada (60.4%), with the remainder from Western Canada (6.9%), the Prairies (25.8%) and Atlantic Canada (6.9%). No cases were reported from Northern Canada. For reporting purposes, the year of diagnoses of a positive laboratory test for HSV was used. The high number of duplicate reports of individual cases (44/122) is indicative of physicians' buy-in to this comprehensive voluntary mechanism of case ascertainment.

The overall demographic and health profile of the 58 confirmed neonatal HSV cases is summarized in Tables 22 and 23, for mothers and infants respectively. Table 22 outlines that the majority of mothers were Caucasian, with a mean age of 27, and that most deliveries were vaginal (75.9%) versus Caesarian (24.1%). Importantly, of the 20 women with available information, 12 had a history of HSV infection. Out of 58 cases, only one mother had intrapartum genital HSV lesions.

Approximately half of the neonatal cases were diagnosed by 11 days old, and 51.7% of these cases were female (Table 23). Of the 48 neonates with known HSV type, 18 (37.5%) had HSV-2 and 30 (62.5%) were infected with HSV-1. Over a quarter of cases (27.6%) were delivered prematurely, although the median Apgar score was nine. The majority of infections were localized (63.8%), with almost 90% of these cases localized to the skin.

TABLE 21

Neonatal herpes simplex virus infection reported cases

Status	2000	2001	2002	2003	Total
Confirmed NHSV*	4	24	17	13	58
Cases pending review	0	0	0	0	0
Did not meet entry criteria†	4	8	7	1	20
Duplicates	2	20	18	4	44
Total	**10**	**52**	**42**	**18**	**122**

* Including fatal cases: 1 (2000), 3 (2001), 3 (2002), 2 (2003) = 9 (Total)

† Excluded due to case definition (17), date of diagnosis prior to October 2000 (3)

TABLE 22

Demographic and health profile of the mothers of neonatal HSV cases diagnosed from October 2000 through September 2003 (n=58)

Mean age (years)	27
Ethnicity:	
• Caucasian	74.1%
• Aboriginal	10.3%
• Black	5.2%
• Other	10.4%
Delivery type:	
• Caesarian	24.1%
• Vaginal	75.9%
History of HSV infection prior to delivery*	60.0%
Presence of intrapartum genital HSV lesions	1.7%
HIV infected	0%

* Information available to paediatricians for 20 mothers

TABLE 23

Demographic and health profile of the neonates diagnosed with HSV from October 2000 through September 2003 (n=58)

Female	51.7%
Mean gestational age (weeks)	37.8
Preterm birth (<37 weeks)	27.6%
Mean birth weight (grams)	2906
Median Apgar score at 5 minutes	9
Median age at laboratory diagnosis (range)	11.8 (0–45) days
HSV type*:	
• HSV-1	62.5%
• HSV-2	37.5%
Classification of HSV infection†:	
• Localized	63.8%
• Disseminated	34.5%

* n=48 as ten surveys were missing information on virus type.

† n=57 as one survey was missing information on nature of infection.

Table 24 outlines a comparison of type of infection and age of diagnosis by infant virus type. For the 47 infants with known virus type and type of infection, localized infections were more likely to be caused by HSV-1, while disseminated infections were more likely to be caused by HSV-2 (p=0.017). For the 46 infants with known virus type and age of diagnosis, 19 of 28 HSV-1 infections (67.9%) were diagnosed after the first week of life, while eight of 18 HSV-2 infections (44.4%) were diagnosed in this time frame (p=0.116).

Of the 56 neonates with known treatment information, 98.2% received treatment, all with acyclovir. In the first two months of life, 14 cases (24.1%) displayed obvious sequelae of infection, including seizures (9), encephalitis (10), microcephaly (2), and blindness (1). The overall case fatality rate was 15.5%, with death more likely to occur among disseminated than localized cases (36.8% vs. 2.8%, p<0.001). Of the nine fatal cases, deaths occurred in infants with the following: known disseminated HSV infection (7), localized infection only, negative CSF results on PCR, and respiratory failure secondary to severe CNS insult listed as cause of death (1), and no information on the nature of the infant's infection available with an autopsy pending (1). All nine infants died within 24 days of birth, with seven cases having dissemination to the CNS, four with dissemination to the liver, and three with dissemination to the lungs. For the eight fatal cases with known virus type, six were typed HSV-2 while only two were typed HSV-1 (p=0.016).

Conclusions

Based on 58 confirmed cases from October 2000 through September 2003, the reported neonatal herpes incidence rate in Canada was 5.9 per 100,000

TABLE 24

Comparison of type of infection and age of diagnosis by virus type

	HSV-1	HSV-2	p-value
Type of infection (n=47)			
• Disseminated	5	9	0.017
• Localized	24	9	
Age of diagnosis (n=46)			
• 0-7 days	9	10	0.166
• >7 days	19	8	

live births. This represents a rate that is closer to that reported by the United Kingdom (two per 100,000 live births) than by the United States (20-50 per 100,000 live births). Over a third of these infections were disseminated cases, with an overall case fatality rate of 15.5%.

The results of this surveillance project have a number of public health implications. Not only is the predominance of neonatal herpes cases attributed to HSV-1 consistent with findings from a growing body of research that indicates an increase in the proportion of genital herpes cases attributed to HSV-1, but the data also shows that herpes vaccine and drug development research for genital herpes needs to account for HSV-1 and HSV-2 genital infections, and demonstrate the effectiveness against both. In addition, study results outline the challenge of preventing neonatal HSV infection and implications for prenatal screening, given that eight out of 20 mothers were unaware of their herpes infection prior to delivery, and obvious genital lesions were present in only one out of 58 cases during the intrapartum period.

As per the project's fifth objective, surviving cases will continue to be followed annually for three years in order to evaluate the overall health and developmental consequences of neonatal HSV infection.

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Neonatal hyperbilirubinemia – severe

(July 2002 to June 2004)

Highlights

- In 2003, 143 cases of newborns affected with severe hyperbilirubinemia were confirmed.
- No etiology was reported in over 70% of cases.
- At presentation, it is essential to perform all laboratory investigations to confirm the etiology.

Background

Even though the occurrence of severe neonatal hyperbilirubinemia and bilirubin encephalopathy is very rare, it can be associated with significant morbidity. Bilirubin encephalopathy is a condition that is unfamiliar to most paediatricians practicing today. In the 1940s and 1950s, bilirubin encephalopathy was a common complication of hyperbilirubinemia associated with Rhesus (Rh) disease and occasionally with ABO hemolytic disease. With the introduction of exchange transfusions and Rh immunoglobulin, a reduction in the occurrence of bilirubin encephalopathy was noted. Also, better antenatal monitoring and availability of intrauterine blood transfusion has eliminated most cases of erythroblastosis fetalis secondary to Rh disease. Phototherapy has drastically reduced the need for exchange transfusions. Despite this, in the last several years, reports of bilirubin encephalopathy associated with extremely high serum bilirubin levels have increased (Penn et al., 1994, MacDonald et al., 1995, Maisels et al., 1995). In most cases, term infants appeared to be healthy and breast-fed with no evidence of obvious hemolytic disease (Rh disease or other antibody-related hemolysis).

Based on epidemiological studies, a number of risk factors have been found to be associated with severe

hyperbilirubinemia in the newborn. These include jaundice presenting in the first 24 hours, jaundice noted at discharge from the hospital, previous sibling with jaundice, gestational age between 35 and 38 weeks, breast-feeding and infant bruising and cephalhematoma (Dennery et al., 2001, Newman et al., 2000). Additional risk factors identified by laboratory investigations include Rh and ABO incompatibility and glucose 6 phosphate dehydrogenase (G6PD) deficiency.

The frequency of severe neonatal hyperbilirubinemia during the current era has not been well documented. Attempts to better quantify its frequency, etiologies and associated risk factors in Canada would be of value prior to identifying strategies for risk reduction. Information obtained from a screening program for the detection of G6PD deficiency or routine determination of blood group and Coombs' analysis on cord blood may help to achieve risk reduction.

Objectives

The main study objective is to obtain epidemiological data on the incidence of severe neonatal hyperbilirubinemia and bilirubin encephalopathy, the burden of illness with regard to medical treatment (phototherapy, transfusions and exchange transfusions), and the neurodevelopmental outcome. Attempts will be made to identify the timing of presentation of jaundice, as well as the etiology and associated triggering or risk factors. This information will help in the development of prevention strategies (G6PD deficiency screening program, cord blood group and Coombs' test and educational programs).

Case definition

Term infants 60 days of age or less with unconjugated hyperbilirubinemia who have had either:

1) Peak serum total bilirubin >425 μmol/L or
2) Neonatal exchange transfusion

Exclusion criteria

Infants who have had exchange transfusion for well-documented Rh isoimmunization disease or are less than 36 weeks gestational age.

Results

Of the 178 cases of severe neonatal hyperbilirubinemia that were reported in 2003, 143 met the criteria for inclusion, eight are still pending review, 19 were duplicates and a further eight cases were discarded. The cause of severe hyperbilirubinemia was identified in less than a third of the cases (42/143). The etiologies included: ABO incompatibility (28), G6PD deficiency (7), other blood group incompatibility (2), urinary tract infections (2), spherocytosis (1), pyuvate kinase deficiency (1) and congenital hypothyroidism (1). The average peak bilirubin reported was 477 μmol/L with a range of 137-773 μmol/L. The infant with a bilirubin level of 137 μmol/L required an early exchange transfusion. Of the 143 confirmed cases, 126 neonates required phototherapy, 33 had an exchange transfusion while seven had transfusions of packed RBC's. Non-standard treatments included IVIG, albumin and phenobarbital.

Conclusions

Severe neonatal hyperbilirubinemia continues to occur in term neonates. In a significant proportion of

TABLE 25

Severe neonatal hyperbilirubinemia reported cases

	Reported	Confirmed	Duplicates	Discarded	Detailed report form not returned
2002	91	60	10	17	4
2003	178	143	19	8	8
Total	**269**	**203**	**29**	**25**	**12**

reported cases, the underlying etiology could not be identified. This is partly attributable to incomplete evaluation of these infants at the time of admission. This finding highlights the importance of a complete hematological workup at the time of presentation, including a CBC, peripheral smear, screen for maternal and infant blood group, Coombs' testing and a G6PD screen. ABO incompatibility and G6PD deficiency were identified as important etiological agents in screened infants. These findings suggest not only that consideration should be given to implementing routine neonatal screening, but also that increased awareness and appropriate feedback to practicing physicians responsible for care of such neonates are needed.

Severe neonatal hyperbilirubinemia and its associated long-term neurological sequelae is a potentially preventable disorder. Several of the cases in this study were neurologically abnormal at the time of presentation. Although this study wasn't designed to assess the incidence of long-term neurological disease, such as kernicterus, a subsequent study addressing questions pertaining to the prevalence of kernicterus and its associated morbidity would be invaluable.

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Prader-Willi syndrome

(January 2003 to December 2004)

Highlights

- Preliminary data results suggest a prevalence of 1:6,700 to 1:10,000.
- As a result of increased awareness in the paediatric community, older PWS patients are being confirmed.
- Diagnosis is now confirmed more often by improved genetic testing than by clinical evaluation.

Background

Prader-Willi syndrome (PWS), an abnormality of chromosome 15, is the most common monogenetic cause of obesity (estimated at 1:10,000 to 1:15,000) with hypothalamic dysfunction leading to hyperphagia and consequent obesity with secondary sequelae of diabetes, heart disease, stroke, sleep apnea, and potential death. Prevalence data in PWS is not well known in Canada, but is drawn from estimates on other geographical locales, or small population studies. Knowing the true Canadian PWS incidence and clinical status on diagnosis will allow for a better understanding of the challenge to be faced and will help with future health-care planning, both on an individual level (intervention strategies) and on a population basis (resource planning), regarding socio-economic support, design and obesity prevention strategy programming.

Objectives

1) To determine the incidence and the mean age of PWS diagnosis in Canada.

2) To ascertain the method of PWS diagnosis: clinical and/or genetic.
3) To create an awareness in the scientific community of PWS.

Case definition

Any child up to and including 18 years of age with newly diagnosed PWS confirmed clinically (PWS clinical score), and/or genetically (methylation and/or FISH [fluorescent *in situ* hybridisation] test). A clinical diagnosis* of PWS relies on a score derived from major and minor criteria:

- < 3 years: 5 points (4 from major criteria)
- > 3 years: 8 points (5 from major criteria)

* *See www.cps.ca/english/cpsp/studies/prader.htm for complete clinical scoring*

Discussion

The PWS incidence is predicted at 22 cases per year. Preliminary data results suggest that so far two-thirds of this predicted number are being identified. Interestingly, the study also identified 17 PWS cases from previous years, providing important comparative data needing further analysis. Additionally, two of the patients confirmed genetically in 2003 were 14 and 39 years of age. As a result, the average age of diagnosis (5.4 years) is high for the 2003 cohort, compared to 2.33 years for confirmed cases diagnosed prior to the initiation of the study. Now the trend in the paediatric community is toward a genetic diagnosis (54.5% in 2003 vs. 42.8% in previous years), which is reducing the age of confirmation of PWS diagnosis to 1.4 years compared to 2.33 years previously. In subsequent years, this average age of diagnosis will undoubtedly go down even further, as more and more children will be diagnosed genetically.

With new genetic testing, confirmation is now more readily available, allowing for earlier diagnosis and treatment. Early establishment of environmental control and behavioural management will make great strides in improving the health and well-being of children and youth affected with Prader-Willi syndrome.

TABLE 26

Prader-Willi syndrome reported cases

PWS 2003	PWS prior years	Duplicates	Discards	Pending	Total
14	17	14	3	8	56

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Vitamin D deficiency rickets

(July 2002 to June 2004)

Highlights

- From July 2002 until December 2003, 69 cases of vitamin D deficiency resulting in rickets were confirmed among children living in Canada.
- The vast majority of confirmed cases were infants and toddlers with intermediate and dark skin, who had been exclusively breast-fed without vitamin D supplementation.
- These results indicate that despite the current Canadian Paediatric Society recommendation that exclusively breast-fed infants receive vitamin D supplementation (400 IU/day; 800 IU/day for infants living north of the 55th latitude), rickets remains a major public health problem in Canada.

Background

Vitamin D is necessary for calcium homeostasis and for mineralization of the growing skeleton. A deficiency in vitamin D during childhood is associated with potentially significant clinical consequences, as it leads to a mineralization defect of the growth plates (rickets) and of bone tissue (osteomalacia). Poor linear growth and skeletal deformity are hallmarks of vitamin D deficiency during childhood, in addition to hypocalcemic seizures, abnormal dentition, and delayed developmental milestones. The disease is entirely preventable through such simple measures as ensuring adequate dietary intake of vitamin D or administration of a daily supplement.

Recent literature has proposed that the incidence of vitamin D deficiency rickets is rising in many countries worldwide, and clinical experiences suggest that Canada may be no exception. This is despite the regulated, Canadian public health policy that all fluid dairy products (excluding yogurt drinks) are fortified with vitamin D, as infants and children living in Canada cannot depend upon adequate skin exposure to sunlight for vitamin D synthesis. Furthermore, the Canadian Paediatric Society (CPS) has recommended that all exclusively breast-fed infants receive a daily supplement of oral vitamin D, since breast milk is not a rich source of this nutrient. Despite these preventive measures, vitamin D deficiency rickets appears all too frequently in Canada, with certain geo-ethnic groups continuing to be at heightened risk for developing the disease. The main purpose of this study has been to determine the incidence of vitamin D deficiency rickets among children living in Canada by seeking reports of all newly diagnosed cases between July 2002 and June 2004.

Objectives

1) To ascertain the incidence of simple vitamin D deficiency (nutritional rickets) among children living in Canada by identifying all newly diagnosed cases over a two-year period.
2) To obtain demographic and medical information to facilitate the identification of children at risk for developing the disease.
3) To evaluate the efficacy of current strategies to prevent the development of the disease in Canada.
4) To supply data that will assist with the development of novel public health policies to prevent nutritional rickets among children living in Canada.

Case definition

Children up to and including 18 years of age with calcipenic rickets secondary to simple vitamin D deficiency (also known as nutritional rickets).

Inclusion criteria

1) Low serum 25-hydroxyvitamin D (25OHD)
2) Elevated serum alkaline phosphatase

Exclusion criteria

1) Vitamin D deficiency rickets associated with underlying disease, such as fat malabsorption, liver disease and renal insufficiency, and with illnesses necessitating total parenteral nutrition.
2) Vitamin D deficiency secondary to heritable disorders of vitamin D metabolism, including:
   - 1α-hydroxylase deficiency (pseudo-vitamin D deficiency rickets, PDDR)
   - vitamin D receptor defects (hypocalcemic vitamin D resistant rickets, HVDRR).
3) Phosphopenic rickets of any etiology (where hypophosphatemia is the primary cause of the rickets, and not due to calcipenic rickets with secondary hyperparathyroidism).

Results

Between July 2002 and December 2003, there were 111 reports of vitamin D deficiency rickets among children living in Canada, of which 69 were confirmed cases, 21 were duplicates, ten were discarded because of failure to meet the inclusion criteria and 11 are still under review. Summaries of the results for 2003 and the cumulative data are presented in Tables 27 and 28.

Demographic data

The majority of confirmed vitamin D deficiency cases (49%, 34/69) were from Ontario, with an additional 16% from Alberta, 13% from Quebec, and the remaining 22% divided among British Columbia, Manitoba, Northwest Territories, Nunavut, Saskatchewan and Nova Scotia. There were no reported cases of rickets from the Yukon, New Brunswick or Prince Edward Island. The gender was indicated for 97% of the cases, with 45% being female and 52% male. The mean age at diagnosis was 1.46 years (0.95 standard deviations, SD; range 0.10-6.34). Sixteen percent (16%) of the cases had immigrated to Canada in the months preceding diagnosis. Thirty-three percent (33%) of the confirmed cases were Black, 14% were First Nations, 13% were Middle Eastern, 10% were Inuit, 9% were Caucasian, and 1% were Asian, while the ancestry was not indicated in the remaining 20% of cases. The mean maternal age at the time of delivery was 29 years (SD 6.3; range 18-39).

Risk factors for vitamin D deficiency

Eighty-six percent (86%) of the confirmed cases were classified as intermediate or dark-skinned. However, fair-skinned children living in Canada were not exempt from developing the disease, as they comprised 12% (8/69) of the confirmed cases. Skin colour was not indicated for 3% of the cases. Fourteen of 71 mothers were veiled during, and following, pregnancy. Physicians reported that 85% of the cases had been breast-fed, while the feeding status was not indicated in the remaining confirmed cases. As expected, the vast majority of the cases (86%, 59/69) had not received vitamin D supplementation prior to the development of the disease. In the remaining 14% of cases, supplementation with vitamin D was either not indicated on the case report form or was unreliably administered by caregivers prior to diagnosis. Only 16% of mothers were documented as having received vitamin D supplementation during pregnancy. Following delivery, this number fell to 6%, and the majority of mothers did not drink milk in the post-natal period. Milk allergies and sunscreen use did not appear to be significant risk factors for vitamin D deficiency among children in Canada, as only three of the confirmed cases manifested a milk allergy, while frequent sunscreen use was documented in only two of the cases.

TABLE 27

Vitamin D deficiency rickets results in 2003

Reported	Confirmed	Duplicates	Excluded	Under review
78	45	16	7	10

TABLE 28

Vitamin D deficiency rickets cumulative results: July 2002 to December 2003

Reported	Confirmed	Duplicates	Excluded	Under review
111	69	21	10	11

Clinical and biochemical features at diagnosis

The most frequent signs and symptoms at diagnosis included: skeletal deformity (46%), seizures (15%), failure to thrive (14%), fractures (10%) and delayed milestones (3%). The remaining 12% of cases presented with an incidental discovery of rickets (i.e., identification of a rachitic thoracic cage at the time of x-ray for a respiratory infection). Twenty-nine percent (29%) of patients presented with a combination of these signs and symptoms. Analysis of the serum biochemical parameters of bone and mineral metabolism prior to initiation of vitamin D therapy revealed a mean alkaline phosphatase level of 1,306 U/L (range 296 to 4,969). A 25-hydroxyvitamin D level prior to treatment was available in 48 of the 69 confirmed cases, giving a mean value of 21.0 nmol/L (SD 14.4; range 1-58).

Conclusions

Over 18 months of this two-year surveillance study, 69 cases of nutritional rickets were confirmed among children, predominantly infants and toddlers, residing in Canada. Intermediate- and dark-skinned children who were breast-fed without vitamin D supplementation were at greatest risk for developing the disease, although cases of rickets among fair-skinned children were also documented. A proportion of the mothers were veiled, and most did not receive vitamin D supplementation following delivery, nor did they ingest milk (thus eliminating a potential dietary source of vitamin D). Significant morbidity was present at diagnosis in all confirmed cases, including limb deformity, seizures, failure to thrive, fractures and delayed developmental milestones.

While breast milk should continue to be advocated as the ideal nutritional source for infants and children, it must be recognized that breast milk is not a rich source of vitamin D. This becomes particularly relevant for infants living in northern countries. In view of our northern latitude, the CPSP has recommended since 1998 that all exclusively breast-fed infants receive supplementation with vitamin D (400 IU/day; 800 IU/day for infants living north of the 55th latitude). However, these results suggest that the CPS position statement regarding vitamin D supplementation has not reached all of the general public and has not been universally implemented by health-care providers. Though it is not surprising that children with the greatest number of risk factors for vitamin D deficiency were the most frequently diagnosed, it is surprising that this condition is being detected in a country with ready access to vitamin D supplementation and a clear recommendation from the country's national paediatric organization for prevention of the disease. Evidently, there is an urgent need for heightened awareness of rickets prevention among health-care providers, and education of the general public is warranted. Studies to evaluate the success of educational interventions are a reasonable next step. Consideration should also be given to the role of financial barriers in ensuring

that vitamin D is administered to all breast-fed infants. In addition, alternate modes of therapy, such as antenatal treatment of vitamin D-deficient mothers to ensure adequate stores during breast-feeding, are worthy of future investigation.

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New Studies in 2004

Acquired demyelinating syndromes of the central nervous system

(April 2004 to March 2007)

"Children experiencing acquired demyelinating syndromes are at risk for the development of multiple sclerosis."

Acquired inflammatory demyelination of the central nervous system (CNS) in childhood is a serious illness characterized by acute loss of vision (optic neuritis), loss of function of the spinal cord (transverse myelitis), encephalopathy and multifocal symptomatology (acute disseminated encephalomyelitis), or varied symptoms dependent on the site of demyelination. Advancing our understanding of acute CNS demyelination in children is of utmost importance given that these children may suffer significant acute and long-term morbidity, and are at risk for the subsequent development of the chronic autoimmune disease, multiple sclerosis (MS).

The study proposes to use the CPSP to gather case-specific data to document the clinical features, epidemiological characteristics, familial autoimmune profile, and the current medical care practices provided to children with acquired demyelinating syndromes. This initiative will provide a measure of the impact of CNS demyelination on Canadian children and aims to enhance care of affected children by increasing awareness among Canadian paediatricians of CNS demyelination, and of MS in particular, facilitating prompt and specialized care for children with this disease.

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Acute rheumatic fever

(April 2004 to March 2007)

"In the current era of evidence-based, judicious use of antibiotics, ongoing surveillance of this now rare but serious condition is crucial."

Acute rheumatic fever is a post-infectious collagen vascular disease affecting the heart, joints and central nervous system. It follows untreated group A streptococcal (GAS) pharyngitis after a latent period of approximately three weeks, but does not occur after other GAS infections, such as skin infection (impetigo). Worldwide, acute rheumatic fever remains the commonest cause of acquired heart disease in children, yet the incidence varies widely from region to region, with the vast majority of cases now occurring in developing countries.

The incidence of acute rheumatic fever in developed countries has decreased dramatically since its last peak in the 1970s, but it has not disappeared and remains an important public health issue. The reason for its decrease is not fully understood. The decline in incidence in the early 20th century had already begun prior to the introduction of effective antimicrobial agents, but common use of penicillin to treat symptomatic sore throat, may have contributed somewhat to the decline. Socioeconomic factors,

such as overcrowding and low income are known to be significant risk factors. The majority of cases of rheumatic fever follow cases of pharyngitis due to specific M serotypes of GAS, most commonly 1, 3, 5, 6, 18, 19 and 24, and spontaneous fluctuation of the prevalence of these serotypes is known to occur.

Rheumatic fever is not a reportable condition in Canada, and in the current era of evidence-based, judicious use of antibiotics, ongoing surveillance of this now rare but serious condition is crucial. Rheumatic heart disease is a lifelong complication of the condition, which can lead to ongoing medical and surgical needs and can interfere with employment, causing significant socioeconomic impact. However, the risk of developing rheumatic fever must be balanced against the risk of encouraging microbial antibiotic resistance, which is a growing problem in all developed nations and carries its own impact. The study will gather Canadian data on the incidence of first-onset rheumatic fever in childhood and on current treatment practices from coast to coast.

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Adverse drug reactions – serious and life-threatening

(January 2004 to December 2005)

"Adverse drug reactions are responsible for 10% of all hospital admissions. Alarmingly, less than 5% are ever reported to regulators."

Adverse drug reactions (ADRs) are increasingly recognized in North America and Europe as an important cause of childhood morbidity and mortality, yet the true incidence of this problem is poorly defined due to the lack of reporting. In the past, health-care systems have relied steadfastly on the idea of voluntary surveillance systems for the identification and reporting of ADRs. The success of these voluntary systems has been poor, with an estimated 95% of all adverse drug reactions never being reported. In January 2004, the CPSP began active surveillance of ADRs with a goal of generating sufficient numbers of case reports to derive meaningful data for the study of serious and life-threatening ADRs in children.

For this project, a serious and life-threatening ADR to the use of prescription, non-prescription, biological (immunoglobulins) products, complementary medicine (including herbals), and radiopharmaceutical products has been defined as a noxious and unintended response to a drug which occurs at any dose and results in (1) patient hospitalization, (2) emergency department visit, (3) prolonged hospitalization, (4) persistent or significant disability or (5) death.

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Osteogenesis imperfecta

(January 2004 to December 2004)

"With the advent of bisphosphonate therapy for the treatment of children with osteogenesis imperfecta (OI), the prognosis for young OI patients has improved dramatically."

Osteogenesis imperfecta (OI) is a congenital disease of bone characterized by low bone mass and bone fragility. Four different types (OI types I–IV) are commonly distinguished on the basis of clinical features and disease severity, and are frequently due to mutations in type I collagen. Recently, three novel forms of OI with distinct clinical features have been identified and named OI types V–VII. The genetic basis of the novel OI forms has not been elucidated. In the past, OI has been associated with significant morbidity as patients frequently suffered tremendous physical limitations due to recurrent fractures. However, with the advent of bisphosphonate therapy for the treatment of children with OI, the prognosis for young OI patients has improved dramatically.

The current incidence of OI, including the novel OI forms, is presently unknown. The primary aim of this study is to identify the number of new cases of OI diagnosed over a one-year period through the Canadian Paediatric Surveillance Program. Additional study goals are to: (1) identify patients in Canada with novel OI forms in order to obtain clinical and genetic information that may ultimately lead to mutation identification, (2) determine whether a geographic distribution of OI in Canada exists, so that regions in need of a local OI intervention program can be identified, and (3) raise physician awareness about OI in general, and the novel forms in particular, so that diagnoses of OI can be made in a timely fashion, and appropriate therapy can be initiated during the critical years of bone growth and development. Education of paediatricians regarding OI may also assist with the differentiation of an abused child from the child with bone fragility due to OI.

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Severe combined immunodeficiency

(April 2004 to March 2006)

"Timely diagnosis of SCID is crucial as early treatment with bone marrow transplant, enzymatic replacement or gene therapy can be lifesaving."

Severe combined immunodeficiency (SCID) is a group of rare genetic disorders characterized by profound abnormalities in the development and function of the T and B lymphocytes and natural killer cells. Infants with SCID usually present in the

first few months of life with a history of infections that are persistent, recurrent, difficult to treat, or caused by unusual infectious agents. Early diagnosis of SCID is critical because the chances for successful treatment by immune reconstitution are highest for infants who have not yet experienced severe opportunistic infections. Without prompt treatment, nearly all will die within the first year of life.

While no Canadian data on the incidence of SCID is available, it would appear that the rate is higher in the Aboriginal population. Information on the incidence of SCID is required to make an evidence-based decision about the risks (notably disseminated BCG infection) vs. the benefits of offering BCG vaccine to First Nations and Inuit children on reserves with endemic tuberculosis.

The objectives of this study are to estimate the incidence of SCID in the general population and in the Aboriginal population of Canada, and to describe the basic demographics, clinical features and outcomes of the Canadian SCID cases.

The CPSP is the best way to achieve these objectives for two main reasons. First, most cases of SCID in Canada will be captured through this active surveillance mechanism, as the majority will be referred to a paediatrician due to the severity of the disease, and second, because it is a prospective program, information such as Aboriginal ancestry, which is often not available through retrospective chart review, can be collected. Moreover, the study will raise the awareness of Canadian physicians regarding the importance of early diagnosis and prompt treatment of SCID cases.

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Survey Question

Lap-belt syndrome

(February 2003)

To verify that the Canadian Paediatric Surveillance Program was the best venue to achieve the lap-belt syndrome study objectives, a one-time survey was conducted to ensure that paediatricians see children with lap-belt syndrome at some point during their hospitalizations. Results of the 648 responses (29%) confirmed that 150 (22%) paediatricians cared for a child involved in a motor vehicle crash while wearing a lap belt or a lap/shoulder belt. When asked how many, they reported from one to 50, with a mean of 2.2. Forty-seven (7%) paediatricians reported that the child was suffering from a lap-belt syndrome including abdominal injuries (42), lumbar spine injuries (23) and spinal cord involvement (9).

When asked which physicians these children would encounter during their hospital stay, 89% believed they would see an emergentologist, 89% a surgeon, 56% an anesthetist, 53% a paediatric intensivist and 75% a paediatrician; 87% of them believed these children would be seen either by a paediatrician or a paediatric intensivist.

Respondents indicated that patients affected with lap-belt syndrome would either receive care within their institution (62%) or be transferred to a paediatric trauma centre (38%).

While only 29% of the surveys were returned, it is noteworthy that 47 paediatricians reported having taken care of children with lap-belt syndrome in the past year and, in fact, may have taken care of more than one child with these kinds of injuries. It is also very encouraging to see that up to 87% of the children would encounter a paediatrician at some point during their hospitalization.

As a result of these encouraging survey results, the CPSP Steering Committee approved a full two-year study on lap-belt syndrome that began in September 2003.

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International Developments

The International Network of Paediatric Surveillance Units (INoPSU), established in 1998, continues to enhance collaboration between national paediatric surveillance units. INoPSU provides a unique opportunity for simultaneous cross-sectional studies of rare diseases in populations with diverse geographic and ethnic characteristics.

Currently worldwide, there are 13 national paediatric surveillance units that are full members of INoPSU: Australia, Britain, Canada, Germany, Ireland, Latvia, Malaysia, Netherlands, New Zealand, Papua New Guinea, Portugal, Switzerland, and Wales. The Cyprus/Greece surveillance unit is an affiliate member until such time as it fulfills the requirements of full membership. As well, the British Ophthalmological Surveillance Unit is an associate member. Argentina and Trinidad and Tobago are currently developing surveillance units.

The first formal INoPSU meeting was held in Ottawa in June 2000 followed by a second meeting in York, England, in April 2002. As a result of these successes, a third INoPSU meeting is planned for Portugal in the spring of 2004.

FIGURE 7

Current INoPSU members

TABLE 29

Studies under surveillance by national paediatric surveillance units in 2003

Abdominal injury due to child abuse	BPSU
Acute encephalitis	PPSU
Acute flaccid paralysis	APSU, CPSP, NZPSU, SPSU
Acute rheumatic fever	SPSU
Adverse effects from complementary or alternative medicine	APSU, WPSU
Alcohol and children	IPSU
Anaphylaxis following food ingestion	APSU
Ataxia	NSCK
Atypical mycobacterial infections	ESPED
Atypical tuberculous infection	NSCK
Autism in children under 5 years	IPSU
CHARGE association/syndrome	CPSP
Childhood conversion disorder	APSU
Complicated pneumonia including empyema	WPSU
Congenital cytomegalovirus infection	APSU, BPSU
Congenital rubella syndrome	APSU, BPSU, CPSP, NZPSU, SPSU
Congenital toxoplasmosis	BPSU
Diabetes mellitus	ESPED, IPSU, LPSU, PPSU
Down syndrome	NSCK
Drug/medication-related adverse events	BPSU
Early-onset eating disorder	APSU, CPSP
Fetal alcohol syndrome	APSU
Foregut & hindgut malformations	NZPSU
Fragile X	IPSU
Hemoglobinopathy	NSCK
Hemolytic uremic syndrome	LPSU, NZPSU, PPSU, SPSU
Hepatitis C virus infection	APSU, CPSP

Hereditary periodic fever syndrome	ESPED
HIV/AIDS	APSU, BPSU, LPSU, NSCK, NZPSU
Hodgkin's lymphoma	LPSU
Hypernatremia	NSCK
Hypophosphatasia	ESPED
Idiopathic nephritic syndrome	NSCK, NZPSU
Idiopathic thrombocytopenic purpura	NSCK
Imported tropical diseases: malaria, schistosomiasis, leishmaniasis	ESPED
Inborn errors of metabolism	NZPSU
Ingestion of lamp oil (intoxications)	ESPED
Inherited hypocalcemic salt-losing tubulopathies/Bartter-like syndromes	ESPED
Insufficient breast-feeding	NSCK
Intussusception	SPSU
Invasive fungal infections in very-low-birth-weight (VLBW) infants	BPSU
Invasive group B streptococcus	ESPED, PPSU
Invasive *Haemophilus influenzae* infections (all types)	ESPED
Juvenile idiopathic arthritis	WPSU
Kawasaki disease	CGPSU, PPSU
Kernicterus	ESPED
Langerhans cell histiocytosis	BPSU
Lap-belt syndrome	CPSP
Leukemia	LPSU
Malaria	NSCK
Medium-chain acyl-CoA dehydrogenase deficiency	NSCK
Meningoencephalitis	PPSU
Munchausen by proxy syndrome	APSU
Necrotizing fasciitis	CPSP
Neonatal herpes simplex virus infection	APSU, CPSP, SPSU
Neonatal hyperbilirubinemia – severe	BPSU, CPSP
Neonatal liver failure/perinatal hemochromatosis	CPSP
Neonatal sinus venous thrombosis	ESPED
Nesidioblastosis	LPSU
Neural tube defects	SPSU
Non-Hodgkin's lymphoma	LPSU
Opsoclonus myoclonus syndrome	IPSU
Pancytopenia	CGPSU
Pertussis	CGPSU
Pneumococcal sepsis/meningitis	ESPED, NZPSU
Prader-Willi syndrome	CPSP
Progressive intellectual and neurological deterioration	BPSU
Prolonged infantile cholestasis	NZPSU
Respiratory syncytial virus (RSV) disease	SPSU
Rett syndrome	APSU
Septo-optic dysplasia	WPSU
Shaken baby syndrome	SPSU
Small bowel insufficiency	NSCK
Splenectomy and hyposplenism	WPSU
Subacute sclerosing panencephalitis and complications	ESPED
Subdural hemorrhage (<2 years)	WPSU
Thrombocytopenia	IPSU
Thrombosis	BPSU
Tick-borne encephalitis	SPSU
Tuberculosis	BPSU, WPSU
Varicella/zoster infection	BPSU, ESPED, SPSU
Vitamin D deficiency rickets	CGSPU, CPSP
Vitamin K deficiency bleeding/hemorrhagic disease of the newborn	APSU, BPSU, NZPSU
West syndrome	CGPSU

Legend:

APSU	Australian Paediatric Surveillance Unit	MPSU	Malaysian Paediatric Surveillance Unit
BPSU	British Paediatric Surveillance Unit	NSCK	Netherlands Paediatric Surveillance Unit
CGPSU	Cyprus/Greece Paediatric Surveillance Unit	NZPSU	New Zealand Paediatric Surveillance Unit
CPSP	Canadian Paediatric Surveillance Program	PNGPSU	Papua New Guinea Paediatric Surveillance Unit
ESPED	German Paediatric Surveillance Unit	PPSU	Portuguese Paediatric Surveillance Unit
IPSU	Irish Paediatric Surveillance Unit	SPSU	Swiss Paediatric Surveillance Unit
LPSU	Latvian Paediatric Surveillance Unit	WPSU	Welsh Paediatric Surveillance Unit

Highlights from other national paediatric surveillance units

Australia

The response of paediatricians and child psychiatrists contributing to the surveillance of mental health issues through the Australian Paediatric Surveillance Unit (APSU) has been very positive. Three psychosocial disorders have recently been monitored: Munchausen by proxy syndrome, conversion disorder and early-onset eating disorder (in children 5-13 years of age).

The Munchausen by proxy syndrome study (2000-2003) has enabled documentation of the presenting features and consequences for a sample of 61 children. The study has also yielded some remarkable information on the impact diagnosing this particular form of child abuse had on the clinician.

Conversion disorder has been described in over 150 children during surveillance through the APSU (2002-2003). Of these children, 74% were female and the mean age was 11.8 years (range=3.0-15.9 years). The clinical picture for most children was complex. Children presented with an average of three symptoms/signs, the most common were motor weakness/paralysis (40%), abnormal gait/movements (39%) and pseudoseizure (25%). Many children also had pain (55%) and/or fatigue (30%). The study has demonstrated that health and support needs of affected children can be very high.

The ongoing study of early-onset eating disorder has confirmed 41 children (73% female) with determined food avoidance to date. Clinical features include weight loss in 88% (mean loss of 6.7 kg), fear of weight gain (76%) and excessive exercise (49%). This study is also currently being conducted through the CPSP, presenting an excellent opportunity for international comparison in the future.

Britain

This past year has seen several new studies come onto our orange card. Of particular importance is the study into childhood tuberculosis (TB). TB notification rates are increasing in the UK, particularly in cities such as London where a quadrupling of notification has occurred over the last ten years. Several factors have contributed to this, including immigration from high prevalence countries and HIV infection. This 13-month study will assess current incidence rates, how children are being managed, what services exist for children with TB, and importantly validate the current national TB surveillance scheme.

In-house, 2003 saw the introduction of the Sir Peter Tizard Research Bursary. This bursary aims to encourage junior doctors to undertake research into rare paediatric disorders and is presented in the name of one of the founders of the BPSU. The successful applicant this year will be examining the epidemiology on thyrotoxicosis.

Cyprus/Greece

The Cyprus/Greece Paediatric Surveillance Unit was established in May 2001 with research projects conducted from April 2002 to December 2003. Cases were reported on West syndrome, Kawasaki disease, rickets, pertussis and pancytopenia. The unit is now applying for full membership at the 2004 INoPSU meeting in Portugal.

Germany

Cerebral venous thrombosis in children was studied to assess the association of prothrombotic risk factors and underlying conditions (infections, vascular trauma, immobilization, malignancies, autoimmune diseases, renal diseases, metabolic disorders, obesity, birth asphyxia, cardiac malformations, and use of

prothrombotic drugs) with cerebral venous thrombosis (CVT) in children.

Results were published in *Circulation* 2003;108: 1362-7 (Cerebral venous thrombosis in children – A multifactorial origin). From 1995 to 2002, 149 paediatric patients aged newborn to <18 years (median six years) with CVT were consecutively enrolled. In patients, and in 149 age- and gender-matched children with similar underlying clinical conditions but without CVT, the factor V G1691A mutation, the factor II G20210A variant, lipoprotein(a) [Lp(a)], protein C, protein S, antithrombin, and antiphospholipid antibodies, as well as associated clinical conditions, were investigated. Eighty-four (56.4%) of the patients had at least one prothrombotic risk factor compared with 31 control children (20.8%; P<0.0001). In addition, 105 (70.5%) of 149 patients with CVT presented with an underlying predisposing condition. On univariate analysis, factor V, protein C, protein S, and elevated Lp(a) were found to be significantly associated with CVT. However, in multivariate analysis, only the combination of a prothrombotic risk factor with an underlying condition (OR 3.9, 95% CI 1.8 to 8.6), increased Lp(a) (OR 4.1, 95% CI 2.0 to 8.7), and protein C deficiency (OR 11.1, 95% CI 1.2 to 104.4) had independent associations with CVT in the children investigated.

CVT in children is a multifactorial disease that, in the majority of cases, results from a combination of prothrombotic risk factors and/or underlying clinical condition.

Ireland

The type I diabetes study is going into its second year and will compare current insulin figures to the previous survey in 1997. A large number of reports have been seen; the impression prior to analysis is that there is a continuing increase in new onset type I diabetes in children as in other countries. The last survey put the Republic of Ireland and Northern Ireland into the moderate to high group of countries concerning diabetes mellitus, and the results of the 2003-2004 study are awaited with interest.

The study on alcohol and children concluded that acute, severe alcohol ingestion by children resulted in hospital admissions and, of concern, produced a high number of adolescents, particularly girls, presenting to children's units in an intoxicated state. The results of this study will be presented at the spring meeting of the Irish Paediatric Association and have already generated some media interest.

Knowing that a similar study is ongoing in the British unit, the IPSU put congenital toxoplasmosis on their card, in order to ascertain as completely as possible the frequency of this condition in Ireland.

Latvia

Latvia continues active surveillance in the following areas: hematology/oncology, endocrinology, nephrology and HIV/AIDS.

Netherlands

Of note in 2003 was the rise in the incidence of hemoglobinopathies compared to the surveillance numbers in 1993. A national screening program for sickle cell disease and thalassemia will be set up. Medium-chain acyl-CoA dehydrogenase deficiency surveillance shows that the incidence is as expected and no children are missed by the tandem mass spectrometry screening in the northern part of the Netherlands where a pilot screening is conducted.

New Zealand

The continuation of a very high response rate, consistently 94 to 96%, demonstrates the interest expressed by paediatricians in using the scheme for their research. It has also helped to secure a further

three years of funding from the New Zealand Ministry of Health.

Portugal

The Portuguese Paediatric Surveillance Unit (PPSU) is a scientific branch of the Portuguese Paediatric Society, with no state institutional links. Based on the British Paediatric Surveillance Unit, the PPSU was created in June 2000 and began active surveillance in April 2001. The population under national surveillance is estimated to be 1,673,600 children and adolescents less than 15 years of age. The PPSU includes on its mailing list all registered paediatricians, paediatric surgeons, neurologists and cardiologists, as well as the residents of these specialties for a total of 1,506 participants. Notification is individual, not institutional. Both postal and electronic notification is available for the return of the postal card and the case inquiry.

During the year 2003, the return rate of notifying cards oscillated between 20 and 25%, amounting to circa 300 cards returned monthly. Of the 3,340 cards returned in 2003, 145 (4.34%) participants reported cases. More than one hundred cards are returned electronically each month. The confirmation of notified cases is not yet closed, as circa 50% of the case inquiries have not yet been returned.

So far, the PPSU has been able to confirm the national dimension of the surveillance system. Data confirm the utility of including non-hospital-based physicians on the mailing list, particularly for some conditions under surveillance such as diabetes. The large mailing list includes every paediatrician (or related specialist) in the country, both active and retired, as well as residents. The PPSU knows that this decision has had a negative effect on the return rates, but feels that it has elements to suggest that reporting has not been negatively affected. This fact supports the decision of keeping every member of the Portuguese Paediatric Society on the mailing list, despite low return rates, at least for the time being.

Switzerland

In July 2002, shaken baby syndrome was included in the SPSU program (under the scientific direction of Dr Ulrich Lips, paediatric university hospital, Zurich). The goal of the study was to ascertain the incidence of children presenting with typical symptoms or "forme-fruste" of shaken baby syndrome and to sensitize the medical profession to this special type of infantile abuse. Unfortunately, in Switzerland, incidence data relating to this syndrome is not available. The public ministry of the Zurich district is the only one (approximately 17% of the Swiss population) that inventoried about ten cases in the years 1999-2001. During this same time frame, the paediatric hospital of Zurich had three children with severe shaken baby syndrome. The number of clinical findings should be higher as the syndrome is not well known. The case definition bases itself on the following points: 1) clinical symptoms (bulging fontanel, convulsions, disturbances, impaired conscience); 2) eye examination signs (unilateral or bilateral retinal and/or vitreous hemorrhages); 3) medical imagery signs tomodensitometry/sub-dural or sub-arachnoidal hematomas parenchymatous lesions (differentiation of gray/white/MRI matter, contusions, shearing injuries, etc.); 4) clinical history (child shaken in the sagittal direction). Depending on the situation, a child deceased as a result of a shaking trauma is not transferred to a clinic but rather directly to a medical legal institution where data collection is carried out simultaneously with these institutions. From July to December 2002, seven shaken baby syndrome cases were reported in children six months old or less, the average age being 5.3 months. Two children succumbed very quickly, another did not present any clinical or radiological sequelae at the time of hospital discharge, and four

had neurological sequelae. In 2003, only five cases were declared. Long-term follow-up is undertaken through a second questionnaire that is sent nine months after the initial trauma.

Wales

The incidence of juvenile idiopathic arthritis based on reporting to the WPSU in the first six months suggests an incidence of five per 100,000 population under the age of 16 years. This is lower than expected and approximately half the incidence rate seen in the previous UK study of hospital referred juvenile arthritis. It is likely that there is under-reporting, but it is also possible that these six months may be unrepresentative and that the incidence may be different when looked at over the intended two years of the survey period. The number of children referred for physiotherapy, occupational therapy and orthotics is lower than expected (ideally all patients with a diagnosis of juvenile arthritis should receive specialist advice from a paediatric physiotherapist experienced in dealing with juvenile arthritis). The numbers of patients receiving disease-modifying drugs and anti-TNF (tumor necrosis factor) treatment is appropriate for a newly diagnosed population.

Call for New Studies

Research opportunities

Wanted

- Investigators to initiate new CPSP studies on childhood disorders that are high in disability, morbidity, mortality and economic costs to society, despite their low frequency.
- The paediatric community to take up the challenge of proposing a wide range of research studies.
- Interested individuals prepared to assume a leadership role in developing protocols and analyzing study data.

The tool

The CPSP is:

- A well-established, timely, cost-effective surveillance infrastructure.
- A multi-faceted surveillance tool capable of collecting reliable data in a variety of different fields.
- An effective means of monitoring low-frequency, high-impact diseases and conditions.

Track record

- An 83% overall initial response from more than 2,300 paediatricians.
- An impressive 96% data completion rate for the 583 cases reported in 2003.
- High duplicate reporting rate (21.3%) assuring case ascertainment and participant commitment.

International flavour

- Be part of INoPSU (International Network of Paediatric Surveillance Units), a growing network of national paediatric surveillance units that exists in 13 countries around the world.
- Take advantage of international collaboration, as INoPSU studies provide a remarkable opportunity to compare similar data and learn more about rare diseases worldwide.

Looking for ideas?

Here are a few examples of studies suggested through the program evaluation survey:

- Abdominal wall defects – gastroschisis
- Brachial plexus injury
- Complications of body piercing
- Congenital varicella
- Heavy metal poisoning
- Histiocytosis disorders
- Laryngeal papillomatosis
- Kawasaki disease
- Severe iron deficiency in preschoolers
- Type 1 diabetes
- Biliary atresia
- Circumcision complications
- Congenital parvovirus B19 infection
- Familial melanoma
- Herpes zoster in children
- Invasive group B streptococcus in neonates
- Neonatal *Listeria* infections
- Severe hypernatremia
- Sudden death in asthma

The potential for new studies in different paediatric subspecialties is endless. If you have a research project in mind, please contact the CPSP Senior Coordinator at 613-526-9397, ext. 239, for more information.


CPSP