Congenital Anomalies Surveillance in Canada

R. Brian Lowry, MD, DSc, FRCPC

ABSTRACT

Congenital anomalies (CA) are present in approximately 3% of all newborn babies and account for about 12% of paediatric hospital admissions. They represent an important public health problem. Surveillance is especially important so that preventive measures such as folic acid fortification can be properly assessed without resorting to a series of ad hoc studies. Canada's surveillance of CAs is weak, with only Alberta and British Columbia having established systems. Most provinces have perinatal systems but their CA data are incomplete and they do not capture terminations of pregnancy. The same is true of the Public Health Agency of Canada's system. A new system, the Fetal Alert Network, has been proposed for Ontario, which represents a start but will require additional sources of ascertainment if it is to be a truly population-based system for Ontario.

Key words: Congenital anomalies; surveillance; Canada; provinces; prevention

RÉSUMÉ

Des anomalies congénitales (AC) sont présentes chez environ 3 % des nouveau-nés et sont la cause d'environ 12 % des hospitalisations en pédiatrie. Elles constituent donc un important problème de santé publique. La surveillance est particulièrement importante, car il faut pouvoir évaluer convenablement les mesures de prévention (comme l'enrichissement des aliments en acide folique) sans recourir à une série d'études spéciales. La surveillance des AC présente des lacunes au Canada, où seules l'Alberta et la Colombie-Britannique ont des systèmes établis à cet effet. La plupart des provinces ont des systèmes de surveillance périnatale, mais leurs données sur les AC sont incomplètes, et les interruptions de grossesse n'y sont pas enregistrées. Il en va de même pour le système de l'Agence de la santé publique du Canada. Un nouveau réseau, le Fetal Alert Network, a été proposé pour l'Ontario; c'est un début, mais pour en faire un système vraiment représentatif de la population ontarienne, il faudra trouver des sources de vérification supplémentaires.

Mots clés : anomalies congénitales; surveillance; Canada; provinces; prévention

Congenital anomalies are present in approximately 3% of all newborns¹ and are the leading cause of infant mortality. They account for about 12% of admissions to pediatric hospitals in the US² and are an important public health problem, hence the importance of good congenital anomaly data. The full impact of congenital anomalies can be measured in years of lost life, medical costs and years of lost income/productivity.

Currently Canada has three Congenital Anomalies Surveillance Systems: the Canadian Congenital Anomalies Surveillance System (CCASS), which is a national system, and two provincial systems – Alberta (AB) and British Columbia (BC). Descriptions of the latter two together with data can be seen in the reports of the International Clearinghouse for Birth Defects Surveillance and Research (www.ICBDSR.org) as both provinces are members who supply data. Further information can be obtained from their websites.*

Most congenital anomalies surveillance systems began after the Thalidomide tragedy of the early 1960s: CCASS and AB in 1966 and BC in 1952. There are essentially three categories of surveillance systems: active, passive with verification, and passive with no verification. In active systems, staff are sent to examine charts, log books, laboratory files, autopsies, etc., whereas passive systems receive the documents from various sources. A further refinement could be the inclusion of matched controls in either an active or a passive system, however there are already considerable privacy issues in having a surveillance system for cases and even more so for controls. Good systems of any variety are improved by: multiple sources of ascertainment; receipt of diagnoses by text and not just by ICD codes; the ability to verify diagnoses and do follow up as required; inclusion of data on terminations of pregnancy (ToPs); and linkage with Vital Statistics regarding live births, stillbirths, and death registrations. Ideally, the inclusion of risk factors is likely to improve research capability.

The Canadian National System

Currently CCASS collects the discharge abstract database (DADs) on newborn

Alberta Congenital Anomalies Surveillance System, Alberta Health & Wellness, Department of Medical Genetics, University of Calgary & Alberta Children's Hospital, Calgary, AB

Correspondence and reprint requests: Dr. R.B. Lowry, Clinical Genetics, Alberta Children's Hospital, 2888 Shaganappi Trail NW, Calgary, AB T3B 6A8, Tel: 403-955-7370, Fax: 403-955-2870, E-mail: brian.lowry@calgaryhealthregion.ca

Acknowledgement: The Alberta Congenital Anomalies Surveillance System is supported by Alberta Health & Wellness.

* www.vs.gov.bc.ca/stats/hsr/;
www.health.alberta.ca/resources/publications/ACASS_2007.pdf (Accessed August 25, 2008)

infants from hospitals in all provinces either through the Canadian Institute for

Health Information (CIHI) or from the MedEcho system in Quebec. Two provincial exceptions are Alberta where the data are from the Alberta Congenital Anomalies Surveillance System (ACASS) and Manitoba where they are sent directly from the Health Department. Because of privacy concerns by CIHI the data, which are already anonymized, are further anonymized by not having the full date of birth or postal codes. Furthermore the data are received in ICD-coded form and not in text, no verification or follow up of cases is possible and terminations of pregnancy are not included. In other words, CCASS is a passive system with no verification capability. Two other problems are the decision to reduce the upper age of ascertainment from one year to 30 days and the removal of gestational age. The latter potentially leads to overestimation of data on patent ductus arteriosus, undescended testes or pulmonary hypoplasia. The ascertainment time limit, as well as the early obstetric discharge, is a further problem in securing diagnoses, particularly for congenital heart disease. Thus with all of the above limitations, the usefulness of CCASS for surveillance and particularly for research is severely limited. The Public Health Agency of Canada (PHAC) has established a Congenital Anomalies Surveillance Network (www.phac-aspc.gc.ca/ccasn-rcsac) and one of its goals is to improve and expand congenital anomaly surveillance in Canada. The PHAC is fully aware of its shortcomings which have been outlined in a number of publications over the past 20 years.³⁻⁶

Other systems in Canada

Apart from BC and Alberta, there are very limited congenital anomaly surveillance systems in Canada, consisting primarily of perinatal systems (Ontario and Nova Scotia) or maternal serum screen systems (Ontario and Manitoba) or medical genetics registries (Newfoundland). Nova Scotia also has a fetal anomaly register. A new innovative program was recently advanced in Ontario called the Fetal Alert Network⁷ (FAN). This is prospective ascertainment of congenital anomalies in fetal life through to outcome. In my view, however, they need to do more, because to have a

system for the province they need to have total population coverage. It is unlikely that every pregnancy will be captured by them. They also need to extend the ascertainment period well beyond the perinatal period, as they say one of their goals is to promote a congenital anomaly system for Ontario. Some of the limitations have been outlined.⁸

Other national systems

National congenital anomalies surveillance systems may not provide accurate enough data for the majority of anomalies. This is true not only for Canada but also for England and Wales. The systems can be reasonably reliable and accurate for obvious anomalies such as cleft lip and palate, but are unreliable for neural tube defects or chromosome anomalies since a high proportion of these fetuses are terminated prior to legal registration. Maintaining high quality data usually requires a limit to the total size of the population to be covered by a register. As a result, larger nations prefer to have regional rather than national registries, networked nationally such as EUROCAT (European Concerted Action on Congenital Anomalies and Twins)⁹ in Europe and the National Birth Defects Prevention Network (NBDPN) in the United States (www.nbdpn.org).

National congenital anomalies systems can be highly reliable in geographically smaller countries with a long history of public health programs such as Finland, Norway and Sweden or in formerly totalitarian states such as Hungary and the Czech Republic. The unreliability of a national system somewhat similar to Canada's has been demonstrated for England and Wales in the National Congenital Anomalies System (NCAS). Misra et al.¹⁰ have shown that the NCAS for England and Wales had a sensitivity of about 33% when compared to two good regional Registries in Trent and Wales. A further study of NCAS by Boyd¹¹ showed that NCAS had non-uniform and incomplete data, with absence of termination data a major shortcoming.

In the United States of America, there is no national system for congenital anomaly prevalence; rather they have developed a network of state systems known as the National Birth Defects Prevention Network (NBDPN) (www.nbdpn.org).

Some of these states have really excellent active or passive systems with verification, which were reviewed in a 2002 document published by Trust for America's Health (www.healthyamericans.org). A study by Boulet et al.¹² assessed the feasibility of using national hospital discharge data (in the US) to estimate the prevalence of selected birth defects. This would be comparable to CCASS. In the US case, they compared it to the prevalence in 11 of the best state systems from the NBDPN and found that, with the exception of common truncus, the hospital discharge survey data were consistently lower than the NBDPN estimates. The Americans have an advantage in the state-supported system because the Centers for Disease Control (CDC) has provided seed money for systems to get started. Nothing like this is available from PHAC. Another disadvantage of discharge databases is that they do not have unique patient identifiers and cannot be used to locate cases for further study or to coordinate services required by infants with a birth defect. As well, the ability to follow up is necessary if unusual clusters of cases are to be investigated. It would appear therefore that, based on the experience of the United States and England and Wales, good regional registries are likely to provide better, more accurate and timely information.

CONCLUSION

Congenital anomalies continue to be a major public health problem. While the etiology of most is unknown, a first step in prevention is knowing how many cases you have and where they are located as well as other epidemiological factors. This is particularly true now with the advent of folic acid fortification and the clear demonstration by De Wals et al.¹³ in a seven-province study of the efficacy of fortification in the prevention of neural tube defects. Recent evidence suggests that folic acid fortification plus multivitamins has a preventive effect on other congenital anomalies – hence the need for good surveillance systems for evaluation rather than having a series of ad hoc studies.

What should be done about CCASS? Can it be fixed or should it be fixed? It is hard to imagine that it can be fixed given the present conditions. Perhaps PHAC

should concentrate on a few provinces or regions of Canada by providing funds to set up reliable and accurate systems that meet the criteria as outlined in a number of sources.¹⁴

REFERENCES

1. Lary JM, Paulozzi LJ. Sex differences in the prevalence of human birth defects: A population based study. *Teratology* 2001;64:237-51.
2. Yoon PW, Olney RS, Khoury MJ, Sappenfield WM, Chavez GF, Taylor D. Contribution of birth defects and genetic diseases to pediatric hospitalizations. A population-based study. *Arch Pediatr Adolesc Med* 1997;151:1096-103.
3. Johnson KC, Rouleau J. Temporal trends in Canadian birth defects birth prevalences 1979-1993. *Can J Public Health* 1997;88(3):169-76.
4. Sherman GJ. The Canadian Congenital Anomalies Surveillance System (CCASS): Four years later. *Chron Dis Can* 1986;6:76-77.
5. Sherman GJ. The Canadian Congenital Anomalies Surveillance System. *Chron Dis Can* 1988;8:58-60.
6. Rouleau J, Arbuckle TE, Johnson KC, Sherman GJ. Description and limitations of the Canadian Congenital Anomalies Surveillance System. *Chron Dis Can* 1995;16:37-42.
7. Kim PCW, Walker M, Beduz M-A. The Fetal Alert Network: An innovative program of access to care, surveillance, and education for birth defects in Ontario. *J Obstet Gynecol Can* 2006;28:1099-102.
8. Lowry RB. The Fetal Alert Network: *J Obstet Gynecol Canada* 2007;29(4):307.
9. Dolk H. EUROCAT: 25 years of European surveillance of congenital anomalies. *Arch Dis Child Fetal Neonatal Ed* 2005;90:355-58.
10. Misra T, Dattani N, Majeed A. Evaluation of the National Congenital Anomaly System in England and Wales. *Arch Dis Child Fetal Neonatal Ed* 2005;90:368-73.
11. Boyd PA, Armstrong B, Dolk H, Botting B, Pattenden S, Abramsky L, et al. Congenital anomaly surveillance in England – ascertainment deficiencies in the national system. *Br Med J* 2005;330:27-31.
12. Boulet SL, Correa-Villasenor A, Hsia J, Atrash H. Feasibility of using national hospital discharge data to estimate the prevalence of selected birth defects. *Birth Defects Res* 2006;Part A 76:757-61.
13. De Wals P, Tairou F, Van Allen MI, Uh S-H, Lowry RB, Sibbald B, et al. Impact of folic acid food fortification on the prevalence of neural tube defects in Canada. *N Engl J Med* 2007;357:135-42.
14. Meyer RE, Sever SE. Towards a gold standard for National prevalence estimates of birth defects. *Birth Defects Res* 2006;Part A 76:770-71.

Received: August 16, 2007

Accepted: April 3, 2008

Since 1910, the Canadian Public Health Association has been Canada's Public Health Leader. CPHA:

- ☒ encourages citizen involvement in Public Health policy and programming;
- ☒ brings together diverse individuals and organizations, creating a united voice on Public Health issues in Canada and around the world; and
- ☒ champions universal and equitable access to the basic conditions necessary to achieve health for all.

CPHA's strength is its members who give us credibility, direction and authority. To continue to be the voice of Public Health, CPHA needs your expertise and support.

Join your voice to ours.

Join CPHA today.

Call us at 613-725-3769 ext. 118,
e-mail us at « membership@cpa.ca »
or visit us on-line at www.cpha.ca