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Nova Scotia Congenital Anomalies Surveillance System

First Report, 1987-2021

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Background

Gaps exist in the nation's capacity to conduct surveillance for congenital anomalies. In 2002, the Canadian Congenital Anomalies Surveillance Network (CCASN) was established

> To support the development and maintenance of high quality population-based surveillance systems of congenital anomalies that will provide information to improve the health of Canadian children and their families.[1](http://www.phac-aspc.gc.ca/ccasn-rcsac/index-eng.php)

The CCASN produced a series of documents regarding the establishment of provincial/territorial surveillance systems for congenital anomalies[2](), coding of congenital anomalies[3](https://health-infobase.canada.ca/congenital-

[1] Public Health Agency of Canada. What is Canadian Congenital Anomalies Surveillance Network (CCASN)

[2] Canadian Congenital Anomalies Surveillance Network. Recommendations concerning the establishment of congenital anomalies surveillance in Canadian jurisdictions.

[3] Canadian Congenital Anomalies Surveillance Network. Coding of fetal anomalies.

anomalies/), and delineating data elements required for minimal and enhanced surveillance for congenital anomalies[4](https://www.phac-aspc.gc.ca/ccasn-rcsac/sac-cas/pdf/cas-eng.pdf).

In Nova Scotia, data regarding congenital anomalies diagnosed at birth or during the birth admission have been collected since 1987 in the Nova Scotia Atlee Perinatal Database (NSAPD) managed by the Reproductive Care Program of Nova Scotia (RCP). The NSAPD collects data about all live born infants and stillbirths for all fetuses delivered $\geq$ 20 weeks gestation, or $\geq$ 500 grams, or live born and all co-multiples of the aforementioned; the NSAPD also captures information about the mothers of the infants/fetuses mentioned above. The NSAPD does not capture data for congenital anomaly cases identified during admissions that occur after 28 days of age. The NSAPD also does not capture data for pregnancies ending in termination.

Since 1992, data regarding congenital anomalies diagnosed antenatally, or postnatally up to the time of discharge from the IWK Health Centre, or on the basis of an autopsy have been captured through the Fetal Anomaly Database (FADB) housed at the Division of Maternal and Fetal Medicine, IWK Health Centre. The IWK is the only obstetrics/paediatric tertiary care centre in the Maritimes; as such, the FADB captures most congenital anomaly cases in Nova Scotia resulting in live births, still births, and terminations. The FADB does not capture congenital anomaly cases diagnosed after the birth admission, many of which are present in other data sources within the province. The FADB also uses an in-house coding system to classify anomalies, a practice that predates the CCASN recommendation to use the Royal College of Paediatrics and Child Health codes (adapted from the International Statistical Classification of Diseases and Related Health Problems, 10th Revision – ICD-10).

SCAN-NS Activity and Report Summary

1. This is the first provincial report on the prevalence of congenital anomalies in Nova Scotia generated by the Surveillance of Congenital Anomalies in Nova Scotia project.
2. The International Classification of Diseases - 10^{th} Edition (ICD-10-CA) has been adopted by the IWK Health Centre as the reporting classification system and SCA-NS uses the Royal College of Paediatrics and Child Health adaptation of ICD-10 as well. The anomalies

[4]Canadian Congenital Anomalies Surveillance Network. Recommended data variables for congenital anomalies surveillance in Canadian provinces & territories.

outlined in this report reflect the "sentinel anomalies list" adopted by the Nova Scotia Congenital Anomaly Advisory Group. Other anomaly data is collected and available upon request.

3. Nova Scotia continues to participate in the Canadian Congenital Anomalies Surveillance System (CCASS), administered by the Maternal and Infant Health Section of PHAC.
4. The numerator data include live births, stillbirths, and also fetal losses < 20 weeks gestation with congenital anomalies. Denominator data include live births and stillbirths only. By including fetal losses in the numerator, the reported rates should be more representative of true congenital anomaly rates. Fetal losses have been ascertained since 1992.
5. Data provided in this report is a compilation of several data Nova Scotia databases.

- Nova Scotia Atlee Perinatal Database (NSAPD).
- Fetal Anomaly Database (FADB).
- Canadian Institute for Health Information Discharge Abstract Database.
- Vital Statistics Database; Service Nova Scotia.
- Medical Services Insurance (MSI) Claims.

6. Data within SCA-NS includes the years $1992 - 2021$ however data for additional years are available for specific inquiries.
7. Congenital anomaly rates have remained relatively stable over the years with fluctuations occurring on a year to year basis and among specific anomalies.

Introduction

The purpose of SCA-NS is to improve Congenital Anomaly case ascertainment, cluster detection, local and national data comparison and reporting capability. Program objectives

- Provide baseline data on occurrence.
- Identify populations at increased risk.
- Monitor changes in occurrence.
- Investigate clusters.
- Refer affected children to services.
- Evaluate prevention programs.

- Create research opportunities.

SCA-NS includes all Nova Scotia residents, with ascertainment from other Maritime Provinces for those cases where the mother delivered elsewhere. Information on cases includes the prenatal period up to one year of age including live births, stillbirths and terminations of pregnancies in which a fetal anomaly has been detected.

Methodology

Case Definitions

A congenital anomaly is an abnormality that is present at birth, even if not diagnosed until months or years later. Most congenital anomalies are present long before the time of birth, some in the embryonic period (up to the end of the seventh week of gestation) and others in the fetal period (eighth week to term). The term "anomaly" covers all the major classes of abnormalities of development, of which there are four major categories as follows:

- **Malformation -** a morphologic defect of an organ, part of an organ or a larger region of the body resulting from an intrinsically abnormal developmental process (e.g., spina bifida, cleft lip and palate).
- **Deformation -** an abnormal form, shape or position of a part of the body caused by mechanical forces (e.g., extrinsic force such as intrauterine constraint causing some forms of clubfoot).
- **Disruption -** a morphologic defect of an organ, part of an organ or a larger region of the body resulting from the extrinsic breakdown of, or an interference with, an originally normal developmental process (e.g., an infection such as rubella or a teratogen such as thalidomide).
- **Dysplasia -** the abnormal organization of cells into tissues and its morphologic result (e.g., Marfan Syndrome, osteogenesis imperfecta).

Other definitions related to pregnancy outcomes for the purposes of this report are as follows:

- **Live birth -** a complete expulsion or extraction from the mother, irrespective of the duration of the pregnancy, of a fetus in which, after expulsion or extraction, there is breathing, beating of the heart, pulsation of the umbilical cord or definite movement of voluntary muscle (Alberta Vital Statistics Annual review, 2000).

- **Stillbirth -** a complete expulsion or extraction from the mother, after at 20 weeks of pregnancy or more or after attaining a weight of 500 grams or more, of a fetus in which, after the expulsion or extraction, there is no breathing, beating of the heart, pulsation of the umbilical cord or unmistakable movement of voluntary muscle (Alberta Vital Statistics Annual review[5]).
- **Gestation -** completed weeks of pregnancy at delivery.
- **Preterm birth (a.k.a premature) -** a birth before 37 weeks of gestation (< 37 weeks).
- **Termination of Pregnancy (ToP) -** for our purposes, includes any pregnancy loss before 20 weeks gestation (< 20 weeks). Most cases are therapeutic terminations for congenital anomalies but spontaneous abortions or intrauterine fetal deaths with fetal anomalies could also be included.

Anomaly definitions are based, for the most part, on those provided by the International Clearinghouse for Birth Defects Surveillance and Research (ICBDSR) and National Birth Defects Prevention Network (NBDPN).

Case Acertainment

An infant can be ascertained at any time up to the first birthday. Multiple ascertainment of the same infant can occur and is encouraged, as this frequently improves the quality and reliability of the data.

As several malformations may occur in the same infant, it is advantageous to allow each to be reported so that groups of associated malformations may be studied. This, however, leads to difficulties since the final tabulations may be reported as total malformations (anomaly rates) or as the total number of malformed infants (case rates). The tables in [Appendix A3] report anomaly rates, which in most cases are similar to case rates (e.g. cleft palate, hypospadias, microcephaly. Whereas with limb anomalies, there can be multiple different limb anomalies in the same infant.

SCA-NS obtains information about infants with congenital anomalies from a variety of independent sources. Acquisition of additional reporting agencies is always a priority since the use of multiple sources of information improves not only the ease but also completeness of ascertainment as well as for verification of the diagnostic data.

[5] Alberta Vital Statistics Annual review

Quality Control Measures and Data Linkage

Data quality is ensured through both a coded linking process and by manual chart audits of unmatched anomaly records. The first step in data quality control involves the linking of disparate fetal/infant records between databases. Linking is based on the matching of several unique variables in each record.

- If two records "pass": this variable matching process, the records are considered link and therefore report anomalies on the same fetus/infant. This allows SCA-NS to create a record that reports anomalies from several databases for the same fetus/infant.
- If there are records that "fail" the automated matching process they are reviewed manually to determine if that information was only found in one data source or if information was entered incorrectly.

Confidentiality and Release of Data

Authority for the Collection, Use, and Disclosure of Personal Information

In Nova Scotia, personal health information is protected by legislation, policies, and professional codes of ethics of healthcare providers. SCA-NS will follow the provisions outlined in the IWK's Privacy of Personal Health Information policy and the RCP Privacy Policy[6] and the Data Access for Research and Planning Policy and Procedures[7]

Data Requests

Data requests will be reviewed by a sub-committee of the Joint Perinatal Epidemiology Research Unit-Population Health Research Unit-RCP Data Access Committee (JDAC). Clinical and technical staff will participate, as will representatives from all stakeholders currently involved in providing data, i.e. Maternal-Fetal Medicine, BIAP, both with a privacy and a technical representative, and other groups such as Cardiology, as the database develops. Disclosure for research purposes also requires approval by a recognized Research Ethics Board and, if approved, a Data Use Agreement. SCA-NS will only consider disclosing individual level data (with or without personal identifiers) following a technical review and approval from a specially-constituted committee.

[6] RCP Privacy Policy
[7] Data Access for Research and Planning Policy and Procedures

Limitations of Data and Analysis

One of the major limitations of any surveillance system is that on its own, the information provided does not allow etiology to be determined. If trends indicate a potential problem, then separate investigative studies need to be done. However, with appropriate approvals in place, it would be possible to conduct linkage studies with other data sources to explore potential causes of specific birth defects. SCA-NS data are collected passively from a variety of sources and the completeness and accuracy of data are largely dependent on reporting and coding of several different organizations. SCA-NS reviews anomalies that have been entered into the database on a regular basis. Detailed studies of some individual anomalies or anomaly groups aid in the assessment and maintenance of the data quality. With intensive review, some cases might be reassigned, re-coded or discarded altogether from the database. This continuing review might explain some discrepancies in the data from earlier reports.

Epidemiological and Statistical Measures

Unless otherwise stated, the birth defect rates presented in this report are calculated using the following formulae:

$$\text{Anomaly (Defect) rate} = 1,000 \times \frac{\text{Number of a particular congenital anomaly among live births + stillbirths + fetal losses}}{\text{Total number of live births and stillbirths}}$$

$$\text{Case rate} = 1,000 \times \frac{\text{Number of individual infants (live or stillborn) or fetuses with} \geq 1 \text{ congenital anomaly}}{\text{Total number of live births and stillbirths}}$$

Confidence intervals (95%) are also included because the rate obtained is actually only a point estimate of the unknown, true population rate. The confidence interval provides information about the precision of the estimate. Thus, the confidence intervals are an estimated range of values within which there is a 95% probability that the true population rate will fall.

Chi Squared Linear Trend Analysis was performed and presented as appropriate.