

## Introduction

The Health Status Registry (HSR) is a comprehensive database of information operated by the B.C. Vital Statistics Agency. Data on individuals with congenital anomalies, other genetic conditions, and selected disabilities and handicapping conditions are collected in order to keep the public and interested professionals informed, respond to research requests, and conduct major research projects. This report provides information on the contents of the HSR and is an update of the 2002 publication *Health Status Registry, Congenital Anomalies – Genetic Defects – Selected Disabilities, British Columbia to 2000*.

This report outlines the history of the Health Status Registry, and then provides information on the Registry's objectives, confidentiality, registration criteria, diagnostic coding, and data collection. A guide to data interpretation and information about the HSR reporting sources completes the general information section of this report. These reporting sources represent the "data capture" points used for information on congenital anomalies, other genetic conditions, and selected disabilities and handicapping conditions. Ascertainment of registrable cases is dependent upon complete and timely reporting by appropriate sources. Counts of diagnoses by source for 1998 to 2002 are provided in a table at the end of this section. The most significant source of HSR data for this report was discharge abstracts from acute care hospitals throughout the province. A previous comprehensive data review indicated possible gaps in reporting to HSR. As a result, a special data request was made and information was obtained for all conditions recorded on 1984-2000 hospital discharge abstracts that met the criteria for registration with HSR.

The next sections of this report provide information on congenital anomalies, starting with an overview at the provincial level. Tables and graphs show trends in the birth incidence rates of selected congenital anomalies over the last decade. This is followed by information on congenital anomalies at the sub provincial level. The Ministry of Health Services divides the province into six health authorities (HA). Each HA consists of three or four health service delivery areas (HSDAs), which are, in turn, comprised of local health areas (LHAs). Assignment of HSR cases to these geographic areas is based on the mother's address at the time she gave birth. A series of tables provides the incidence of congenital anomalies for single years of birth for each HA and HSDA. Then several years of data are combined in tables showing counts and rates of selected congenital anomaly categories across the areas. The information on congenital anomalies concludes with tables and maps providing statistics by local health area.

The last two sections of this report provide information on "other" genetic conditions and selected disabilities and handicaps as of year-end 2002. Reporting of genetic conditions is not age limited; so these counts are shown for all age groups in the provincial population. Prevalence counts for selected disabilities and handicapping conditions are presented for five age groupings in the provincial child population. Because the purpose of these sections is to monitor where people with disabilities are currently living, all prevalence information is presented by HA and HSDA based upon last known residence address. The report concludes with an appendix listing the diagnostic categories and codes used in the statistical sections of the report, and tables providing birth defects monitoring information for the last three years.

The British Columbia Vital Statistics Agency gratefully acknowledges the input and financial support of the British Columbia Ministry of Children and Family Development.

## History of the Health Status Registry

In 1948, federal government grants were offered to all provinces to establish programs for the prevention, control, and treatment of crippling conditions in children. In British Columbia, the grant was used to determine the number and location of crippled children, the types of crippling diseases, and the availability of treatment facilities. A survey conducted under the auspices of the British Columbia Medical Association was sent to all physicians in the province. The response, however, was very poor. As an alternative, surveys were completed based upon a review of all provincial hospital admission records for 1948 and 1949. Surveys were also sent to public health nurses in the Provincial Public Health Districts.

The completed surveys were sent to the Division of Vital Statistics for tabulation. The survey results found 20,000 crippled children and minimal treatment facilities. When preliminary results indicated that most children were not receiving treatment early enough for maximum benefits, Vital Statistics revised the Physician's Notice of Birth to collect information on abnormalities at birth.

In 1952, the *Crippled Children's Registry* was established under the Division of Vital Statistics in the Ministry of Health and Welfare in order to collect and collate data on crippled children, to assist in the organization of services for these children, and to refer crippled children to appropriate treatment facilities. The criteria for registration were that the individual be less than 21 years of age with a disability severe enough to interfere with normal living, obtaining an education, and earning a livelihood.

In addition to collecting data, this voluntary registry conducted medical examinations to register new cases and reassess children already registered and to refer children to appropriate treatment facilities. Information was provided by Public Health Districts to ensure that cases in their catchment areas received adequate care. The Registry also assisted in the development of services for the handicapped and as volunteer and government organizations were organized, they joined private physicians and treatment and rehabilitation facilities as reporting sources. Mental health centres were also added as reporting sources. The Registry's name was changed to the *Handicapped Children's Registry* in 1957. At this time, there were less than 12,000 cases registered.

In response to the thalidomide disaster in 1960, the age limit of 21 was removed and the name was changed to the *Registry for Handicapped Children and Adults*. The criteria for registration were also expanded to include all familial conditions and congenital malformations. During this decade, more reporting sources were added, including UBC Medical Genetics, death and stillbirth registrations, obstetrical discharge summaries, the Cancer Registry, and hospital admissions/separations for children with congenital anomalies who were less than seven years old. A study to assess the results of treatment and rehabilitation was initiated through the Public Health School Programs. The "7 and 14 year Follow-Up Study" provided information on children between these ages and continued for ten years. During the 1960s the Registry's focus began to change and it became a more passive data repository. By 1968 there were 36,000 cases in the Registry.

In 1975, the Registry's name was changed to the *Health Surveillance Registry* as risk registers for amniocentesis, rubella, hyaline membrane disease, and fetal alcohol spectrum disorder were added. The Registry's objectives were to provide a reliable basis for estimating the prevalence, incidence, and distribution of specific conditions by ascertaining, recording, and classifying handicapping conditions, congenital anomalies, and genetic defects in the population of British Columbia, to assist health services and physicians in the ascertainment, diagnosis, monitoring, and management of such conditions, and to assist medical and genetic research based on the data by undertaking statistical analysis of the data collected. There were approximately 55,000 cases in the Registry in 1973, and almost 100,000 by the end of 1978, largely as a result of an increase in reporting sources.

In 1980, the Cancer Registry, previously part of the Health Surveillance Registry, moved to the Cancer Control Agency of British Columbia. At this time, there were 68 active reporting sources for the HSR and about 108,000 cases. The Registry maintained a low profile during the 1980s amid the provincial government's downsizing and "privatization". By 1989 the Registry had grown to 195,000 cases and more than 80 reporting sources. Beginning in 1975 the HSR Annual Report was replaced by summary tables presented in the Division of Vital Statistics Annual Reports.

In the early 1990s an evaluation of the Registry was conducted and a development plan was prepared which provided recommendations on data collection and recording, access and security, medical coding systems and consultation needs, and technological requirements. In 1991, the *Royal Commission Report on Health Care and Costs* contained a recommendation that Vital Statistics develop and maintain a registry of individuals with disabilities to assist in the development of long-range plans and to monitor the changing needs of the population. Subsequently, in September 1992, amendments to the *Health Act* established the legislative mandate and responsibilities for the HSR.

The Registry's current name (Health Status Registry) was acquired in 1992. In order to refocus the Registry's emphasis on children, the criteria for registration of individuals with long-term physical, mental and/or emotional problems was restricted to persons under the age of 20 years old. Registration of persons with genetic conditions, however, was not age limited. By 2000 there were approximately 215,000 records in the Registry.

In 1998 the Registry started to collect information on early terminations of pregnancy due to congenital anomalies, as recommended by medical geneticists and the B.C. Provincial Health Officer (see *A Report on the Health of British Columbians*; Provincial Health Officer's Annual Report, 1997). Currently a formal agreement is being set up with the participating bodies in order to monitor these cases and to establish reporting procedures in accordance with the *Freedom of Information and Protection of Privacy Act* and legislative mandate.

In 2001 the Health Status Registry was accepted as a full member of the International Clearinghouse for Birth Defects Monitoring Systems (ICBDMS). HSR now participates in ICBDMS's international birth defects monitoring activities by submitting quarterly and annual data for residents of British Columbia to the ICBDMS. In 2002 Autism Spectrum Disorder was added to the Health Status Registry's list of registrable conditions.

## General Information

### ***HSR Mission, Mandate and General Objectives***

The British Columbia Vital Statistics Agency administers section 10 of the Health Act by operating the Health Status Registry. The mission, mandate, and general objectives of the Registry are:

- To record and classify information concerning congenital anomalies, genetic conditions, and selected handicapping conditions of children;
- To assist health care planning and others in the planning and development of appropriate services by providing accurate and reliable data on congenital anomalies, genetic conditions, and chronic handicapping conditions; and
- To undertake statistical analysis of the data collected and to assist medical and genetic research based on this data.

Additional objectives are:

- To keep the public informed by producing timely and accurate statistical-related products, while maintaining the confidentiality of the data;
- To respond to research requests from a wide and varied audience, such as governments, universities, private and public organizations, health-related researchers and planners, and administrators;
- To develop the HSR as a useful tool that will be utilized by health care and social service systems.

### ***Confidentiality***

Maintenance of the confidentiality of HSR data is essential. Confidentiality of the data is protected by ensuring that all staff involved with HSR records comply with the *Freedom of Information and Protection of Privacy Act*, the *Health Act*, and HSR policies and procedures. The *Freedom of Information and Protection of Privacy Act* states that disclosure of personal information is an unreasonable invasion of a personal privacy if the information relates to a medical, psychiatric, or psychological history, diagnosis, condition, treatment, or evaluation. The *Health Act* specifies that persons acting on behalf of the Health Status Registry must not communicate personal identifying information to persons who are not acting on behalf of the Registry except to the extent necessary for recording and classifying information concerning congenital anomalies, genetic conditions, or chronic handicapping conditions. HSR policies and procedures require staff to get prior authorization from the Chief Executive Officer of the British Columbia Vital Statistics Agency before contacting a registered individual or their physician. Registry staff may contact reporting sources concerning registered individuals to clarify information received from the source or to request additional information about the diagnosis or treatment.

Confidentiality is also considered when personal identifiers are not involved. The content of statistical tables is reviewed to ensure small numbers do not allow identification of specific individuals or institutions, and approval for release by the Chief Executive Officer of the British Columbia Vital Statistics Agency or the Director of Information Resource Management is required. The Chief Executive Officer of the British Columbia Vital Statistics Agency may also authorize the release of records without personal identifiers. In order for this to happen, the researcher must first submit a copy of the research proposal and then sign an agreement specifying that the data supplied will only be used for the approved project and that all files will be destroyed upon completion of the project. Data elements that are not personal identifiers but which could allow for the identification of an individual (such as a postal code) are not released.

## Criteria for Registration

Since January 1, 1993, the criteria for registration of a case/individual within the HSR is as follows:

- A person who is 19 years of age or less and is diagnosed as having a physical, mental, and/or emotional problem which is likely to have long-term disabling effects, to interfere substantially with education, or to prevent full and open functional employment; or
- A person of any age who is diagnosed as having a genetic condition or congenital anomaly that is not necessarily disabling.

## Medical Coding

Reporting sources provide identifying and medical information to the Health Status Registry about individuals known to them who meet the criteria for registration. These individuals may have more than one registrable condition/diagnosis. For some cases, a six-digit code based on McKusick's classification system of inherited conditions provides genetic researchers with the specific detail they require to distinguish various genetic conditions and congenital anomalies.

Diagnoses are coded using the World Health Organization's *International Classification of Diseases* (ICD). Coding by HSR staff according to the tenth revision (ICD-10) began in the year 2000, although some reporting sources continued to provide diagnoses using the ninth revision (ICD-9). ICD-10 includes new disease entities and is far more detailed than ICD-9, with approximately twice as many codes. In addition, some groupings of conditions within the classification have changed, and some conditions have been assigned to different parts of the classification than in ICD-9. Although diagnoses from 1999 and earlier years have not been recoded, the categories used for congenital anomalies, other genetic conditions, and disabilities and handicapping conditions presented in this report are based on ICD-10. As a result, some categories presented in this report differ from those shown in earlier reports. More information on the diagnoses used in this report can be found at the beginning of each section, and lists of the ICD-9 and ICD-10 codes used for each category can be found in the appendix.

The HSR has a Medical Policy Committee to provide advice in all medical matters related to the Registry. Members of this committee are appointed by the Assistant Deputy Minister of Knowledge Management and Technology or the Chief Executive Officer of the British Columbia Vital Statistics Agency and currently include geneticists, HSR staff, and Vital Statistics managers. The committee meets on a regular basis, about four times per year. In 2004, the Medical Policy Committee reviewed and revised all HSR registrable conditions and has been working to identify and recruit additional reporting sources.

## Data Collection/Updating

The daily operation of the Health Status Registry includes registration of new cases and updating existing cases. This process begins when information is received from reporting sources. Some sources, such as Children's Hospital, UBC and Victoria General Hospital Medical Genetics, provide information electronically, while others provide hard copies. The information is reviewed to ensure that the case meets the criteria for registration and that the affected person lives in the province. After the medical coding is complete, a search determines if this is an initial registration or if the case is already in the Health Status Registry and data entry is done.

Periodic linkage with Vital Statistics' death files ensures that the death "flag" on HSR records is up to date for individuals who died in British Columbia. This allows the Registry to produce tables of still living cases, as well as snapshots of registrable conditions based on years of birth or at particular points in time. Records for individuals born in the province are also linked to the Vital Statistics' birth files so data pertaining to the individual's birth can be added to their HSR record.

## Information Box

### *Multiple Congenital Anomalies*

The International Centre for Birth Defects (ICBD) monitors congenital malformations by collecting and analysing data from the International Clearinghouse for Birth Defects Monitoring Systems. The Health Status Registry (HSR) provides data to ICBD for dissemination in quarterly and annual reports; an example is provided below.

#### **Cases of Multiple Congenital Anomalies (MCA) Review by Program and Number of Defects 2nd Quarter, 2004**

Program	Total Births	Known Etiology (Syndromes)	1 Major Unrelated Defect	2+ Defects (MCA)	Total Cases Reported
Canada: British Columbia	10,372	3	15	9	27
Finland	14,498	14	1	22	37
France: Central East	27,673	1	4	12	17
Israel: IBDMS	8,654	0	2	5	7
Mexico: RYVEMCE	8,317	2	4	10	16
USA: Atlanta	12,608	1	0	2	3
<b>TOTAL</b>	<b>82,122</b>	<b>21</b>	<b>26</b>	<b>60</b>	<b>107</b>

MCA - Multiple Congenital Anomalies: the current case definition is at least 2 major congenital anomalies (excluding syndromes with known etiology).

Source: International Centre for Birth Defects, 2nd Quarterly Report, 2004.

Please see the Information Box on Collaborative Research by the Health Status Registry for details of other collaborative research studies.

### *Fetal Alcohol Spectrum Disorder*

Diagnosing FASD is difficult because no two individuals with FASD present with the same constellation of anomalies and disabilities. As a result, a diagnosis of FASD requires a referral to a specialist, generally a developmental pediatrician. The HSR has been active within the province of British Columbia in developing the capacity within the province for diagnosing FASD. The Vital Statistics Agency, which operates the HSR, funded a project to develop a plan for training a network of clinicians across the province who can provide the required investigation. The result of this work is the funding proposal *A Plan for Province-Wide Access to Consistent Diagnosis of FAS*. External funding is currently being sought to begin implementation of this plan.

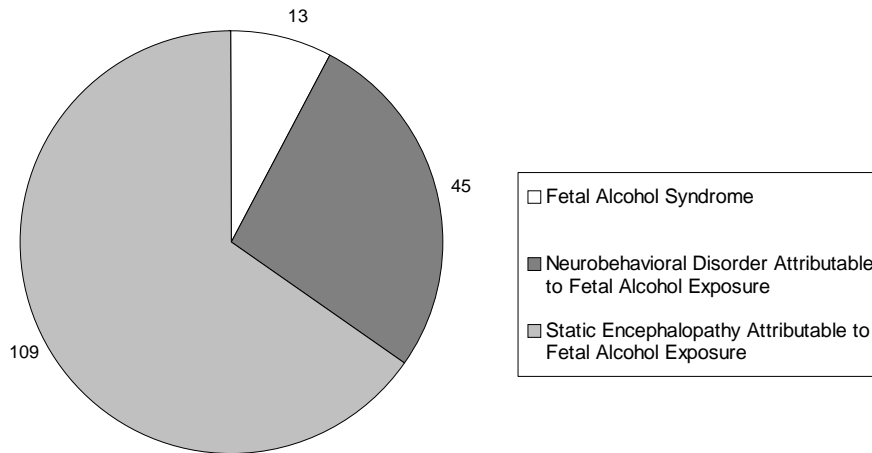
The major reporting sources for FASD in British Columbia are the Asante Centre for Fetal Alcohol Syndrome and Sunny Hill Health Centre for Children. Between 2001 and 2003, HSR received reports of 167 cases with FASD.

The pie chart shows FASD cases by diagnostic category. Cases with Fetal Alcohol Syndrome (FAS) have growth deficiency, specific facial characteristics, and brain dysfunction. The category of Static Encephalopathy Attributable to Fetal Alcohol Exposure consists of cases with detectable brain abnormalities, while Neurobehavioural Disorder Attributable to Fetal Alcohol Exposure consists of cases with cognitive or behavioural dysfunction where brain abnormalities were not detected. Fetal alcohol exposure could be either known or unknown, but not ruled out, for any of the diagnostic categories. Nearly two thirds (65.3%) of the cases reported to HSR have Static Encephalopathy Attributable to Fetal Alcohol Exposure (109 cases). Another quarter (26.9%) of the cases, have Neurobehavioural Disorder Attributable to Fetal Alcohol Exposure (45 cases), while less than one in thirteen (7.8%) cases (13) are FAS.

## Information Box

### *Fetal Alcohol Spectrum Disorder cont'd*

#### Fetal Alcohol Spectrum Disorder Cases Reported to the Health Status Registry



The loss in human potential and cost of caring for children with FASD is very large. To address this issue, the provincial government has developed a comprehensive strategic plan for Fetal Alcohol Spectrum Disorder. It can be found at: <http://www.mcf.gov.bc.ca/fasd/index.htm>.

### **Guide to Data Interpretation**

There are several points to keep in mind when interpreting the kind of information presented in this report. Incomplete case reporting, variations in reporting over time and by areas, and changes in technology have all had an impact upon the HSR. For many of the diagnostic conditions presented in this report, the data should be interpreted as a summary of what is currently available in the Health Status Registry, not as complete statistics on the birth incidence or prevalence of the condition. The remainder of this section elaborates further upon these points.

Some of the rates presented in this report, such as total diagnoses, are based on counts of infants born with any congenital anomaly. Anomalies can vary widely in their severity and in their impact on the future health of individuals afflicted. Some congenital anomalies may not require ongoing health care management or treatment. Therefore, when assessing rates, consideration should be given to the types of anomalies that are contributing to the rates presented in the table.

Tables in this report may show congenital anomaly rates for single years or at the health service delivery area (HSDA) or local health area (LHA) level. For some conditions or for some of the areas, the rates are based on small counts so yearly fluctuations of a few cases will have a large impact. In addition, the completeness of the data may vary depending on the consistency and compliance of reporting by sources.

The reporting sources determine the degree of completeness of registrations and the currency of data on individual cases. HSR case coverage varies by condition, from good coverage for major birth defects that are readily identifiable at birth, to limited coverage for conditions that are usually not diagnosed until late childhood. The time required for diagnosis and reporting also varies by condition, so recent years may have lower counts than earlier years. The reader should always keep in mind that the numbers in this report represent cases or diagnoses reported to the Health Status Registry, not necessarily the number of cases or conditions occurring in each year.

The time span covered by this report has included several changes of emphasis on the reporting cases to the HSR. During the late 1980s and early 1990s some important reporting sources stopped providing data to the HSR, however, since the mid 1990s this situation has reversed and reporting has improved. In 2004 a large amount of missing data was recovered from 1984-2000 hospital discharge abstracts noting conditions that met the criteria for registration with HSR. Nevertheless, the reader is reminded that the long-term trends shown in this document reflect trends in the rate of reporting, as well as trends in the underlying number of cases with the specified conditions in the population.

When comparing HSR data for various areas, the reader is also reminded that the utilization of specialized services may not be uniform across all areas. If residents who live close to a specialized service are more likely to use the service than individuals who live much farther away, HSR data will reflect these utilization rates. Thus higher rates in one area might indicate more services, more utilization of services, or differences in the incidence of various conditions compared to other areas.

Changes in prenatal screening have also had an impact on the HSR data. With the advent of amniocentesis and ultrasound, many types of anomalies can be detected early in pregnancy and the mother may choose to terminate the pregnancy. HSR data on congenital anomalies includes cases that meet the criteria for registration as a stillbirth (a minimum of 20 weeks gestation or a weight of at least 500 grams). Although HSR also collects information on medical terminations of pregnancy (a minimum of 14 weeks gestation) due to congenital anomalies (see the section on the history of the Health Status Registry and Information Box for more details), these cases are not included in the data presented in this report.

Finally, unexplained clusters of cases have often been found when examining congenital anomaly rates over time. Many congenital anomalies show large variations from year to year, even when the rates are stable over longer periods of time. Thus it is quite common to see no cases with a specified anomaly for a number of years, followed by several cases in one year, then fewer or none, and then the pattern repeats.

## HSR Reporting Sources

### *Introduction*

The Health Status Registry reporting sources represent the “data capture” points used for information on congenital anomalies, other genetic conditions, and selected disabilities and handicapping conditions. Ascertainment of registrable cases is dependent upon complete and timely reporting by appropriate sources. The numbers and kinds of reporting sources have changed considerably over time. Although legislation now requires individuals and organizations to submit data to the Health Status Registry, these data have historically been collected from voluntary reporting sources. When the focus of the HSR changed to emphasize registering disabling or handicapping conditions only when they involved children, a number of reporting sources for non-childhood conditions were discontinued. Reported numbers also decreased from some other sources.

Congenital anomalies are present at birth, but only those that are clinically apparent or manifest severe symptoms are likely to be noted on the Notice of Live Birth or Stillbirth, which is completed and sent to the British Columbia Vital Statistics Agency within 48 hours of the birth. Reporting of less evident congenital anomalies, such as pyloric stenosis and many heart defects, depends on contact by the individual with one of the Registry’s other reporting sources such as the British Columbia’s Children’s Hospital, other acute care hospitals, or the HSDA. Large portions of genetic and disabling or handicapping conditions are even less likely to be identified at birth. Cases with genetic conditions are most often reported by UBC and Victoria General Hospital Medical Genetics. For conditions other than those that are present at birth, consistent and complete reporting from sources other than Vital Statistics is essential.

The concept of building case histories through data from multiple reporting sources over an extended period of time is an integral part of this kind of database. Reports on the same individual from several different sources serve to add, update, and correct the existing information, thus improving its accuracy and relevance.

## Highlights

Table 1 shows that a total of 156,389 diagnoses were reported from 1998 to 2002.

The most significant source of HSR data for this report was discharge abstracts from acute care hospitals throughout the province. A previous comprehensive data review indicated possible gaps in reporting to HSR. As a result, a special data request was made and information was obtained for all conditions recorded on 1984-2000 hospital discharge abstracts that met the criteria for registration with HSR. This resulted in a total of 114,857 diagnoses from this source, or nearly three quarters (73.4%) of all diagnoses reported to HSR in 1998-2002. It should be noted that cases previously reported by British Columbia's Women's Hospital and Health Centre, British Columbia's Children's Hospital, or Sunny Hill Health Centre for Children would not be counted in the 114,857 diagnoses from hospital discharge abstracts unless they had additional or different diagnoses than the ones that were previously reported.

There were 41,532 diagnoses from reporting sources other than the hospital discharge data base. Most of these (56.3%) were reported by Children's Hospital. This source reported 23,398 diagnoses.

Vital Statistics Live Births, Stillbirths, Deaths, and Amendments were the source of 12,527 diagnoses, or almost a third (30.2%) of the diagnoses from sources other than the hospital discharge data base.

Table 1  
Number of Diagnoses Provided by Reporting Source  
British Columbia, 1998 - 2002

Reporting Source	Number of Diagnoses
001-040 Public Health	2,317
046 UBC Genetics	1,037
052 Children's Hospital	23,398
060 Canadian National Institute For The Blind	13
064 Vital Statistics Livebirths	9,854
066 Riverview Hospital	5
080 Hospital Discharge Abstracts	114,857
081 Vital Statistics Stillbirths	1,355
082 Vital Statistics Deaths	1,308
097 BC Women's Hospital	340
101 Victoria Medical Genetics	115
167 Children's Hospital Cystic Fibrosis Clinic	51
169 Vital Statistics Amendments	10
170 Willow Clinic - Formerly Woodlands	13
301 Sunny Hill Health Centre For Children	123
401 Asante Centre	22
<b>Total</b>	<b>156,389</b>

Note: Counts include all unique diagnoses provided by the specified reporting source.