Alberta Congenital Anomalies Surveillance System

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The Alberta Congenital Anomalies Surveillance System was started in 1966 in response to the thalidomide tragedy earlier in the decade. It was one of four provincial surveillance systems on which the federal government relied for baseline statistics of congenital anomalies. The government now collects data from six provinces and one territory. The Alberta Congenital Anomaly Surveillance System originally depended on three types of notification to the Division of Vital Statistics, Department of Health, Government of Alberta: birth notice and certificates of death and stillbirth; increased sources of ascertainment have greatly improved data quality. We present the data for 1980–86 and compare the prevalence rates of selected anomalies with the rates from three other surveillance systems. Surveillance systems do not guarantee that a new teratogen will be detected, but they are extremely valuable for testing hypotheses regarding causation. At the very least they provide baseline data with which to compare any deviation or trend. For many, if not most, congenital anomalies total prevention is not possible; however, surveillance systems can be used to measure progress in prevention.

Le réseau albertain de surveillance des malformations congénitales a vu le jour en 1966 en réponse au drame de la thalidomide survenu au début de cette décennie. C'est un des quatre réseaux provinciaux sur lesquels le gouvernement fédéral comptait pour établir des statistiques de base sur les malformations congénitales; celui-ci dispose maintenant de données fournies par six provinces et un territoire. À l'origine, le réseau albertain se fondait sur les déclarations de naissances, de mortinaissances et de décès faites à la Direction des statistiques vitales du ministère provincial de la Santé. Aujourd'hui on a recours à des sources supplémentaires de renseignements, ce qui améliore beaucoup la qualité des données. On présente ici celles des années 1980–86 et compare la fréquence de l'existence de certaines malformations choisies avec ce qu'ont rapporté trois autres réseaux. Si de tels réseaux n'assurent pas la découverte de nouveaux tératogènes, ils sont très utiles pour éprouver des hypothèses étiologiques. À tout le moins ils fournissent des données de base permettant de mettre en évidence toute déviation et toute tendance. Il est impossible de prévenir complètement un grand nombre, sinon la plupart, des malformations congénitales; mais la surveillance permet de juger des progrès dans cette direction.

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The Alberta Congenital Anomalies Surveillance System was started in 1966 as part of the Canadian Congenital Anomaly Surveillance System[1-3] in response to the birth of many infants with severe limb defects due to the perinatal use of thalidomide from 1958 to 1962. The Canadian system now collects data from Alberta, Manitoba, Ontario, New Brunswick, Nova Scotia, Prince Edward Island and the Northwest Territories. Saskatchewan, Quebec, Newfoundland and the Yukon Territory have never submitted data, and British Columbia has stopped submitting data.

The Alberta Congenital Anomaly Surveillance System is the responsibility of the Division of Vital Statistics, Department of Health, Government of Alberta. We describe several key components of this system and present the results of the analyses on the 1980–86 data. The statistics were tabulated for the whole province and for individual health units, and the prevalence rates for selected anomalies were compared with the rates from other jurisdictions.

Ascertainment of data

A birth defect can be ascertained at any time

up to the infant's first birthday. Multiple reports of the same defect can occur and, indeed, are encouraged, because they improve the quality and reliability of the data. The sources of data include birth notices and certificates of death and stillbirth (Table I). The most important source is the Congenital Anomaly Reporting Form, which is forwarded from hospitals and special outpatient clinics to Vital Statistics for record validation and linkage to the birth or death registration. All hospitals in Alberta must submit a Congenital Anomaly Reporting Form on any inpatient under 1 year of age. Anomalies are coded, by means of the British Paediatric Association's adaptation of the International Classification of Diseases ninth revision.[4] Forms with a query are shown to the medical consultant (R.B.L.), who usually writes to the responsible physician for more information. Replies are received in approximately 99% of the cases. If the infant died or was stillborn the registration form will state whether an autopsy was performed; if one was, a copy of the report is obtained. The information is returned to Vital Statistics for data entry and storage on a tape, which is then forwarded to the Canadian Congenital Anomaly Surveillance System.

Not included in the case count are premature infants (born up to 36 weeks' gestation) with one or more of the following anomalies: undescended testis, patent ductus arteriosus and patent foramen ovale.

The data are handled only by Vital Statistics staff. The names of the infants are not revealed, and the staff of the surveillance system and Vital Statistics do not communicate directly with the parents. If contact is necessary, then ethical clearance is obtained through the family physician or community health nurse, depending on the source of the case.

Results

The overall frequency of congenital anomalies in Alberta from 1980 to 1986 was 38 per 1000 total (live and still) births (a rate of almost 4%). However, many of these defects were minor, and the rate of major anomalies was approximately 2.5%.

The prevalence rates of congenital anomalies for individual health units (Fig. 1) varied greatly, from 14.72 per 1000 total births in Fort McMurray to 49.02 in Calgary (Table II). Health units with small numbers of births had much wider 95% confidence intervals than those with large numbers of births. In general the rates were very close to the provincial average. Of all the infants born with defects 57% were boys, and 21% had two or more congenital anomalies.

Table I — Sources of data for the Alberta Congenital Anomalies Surveillance System

Inpatient
Birth notice
Death certificate
Certificate of stillbirth
Congenital Anomaly Reporting Form*
Outpatient
Alberta Children's Hospital (special treatment clinics)
Calgary Biochemical Genetics Laboratory
Calgary Cytogenetics Laboratory
Calgary Medical Genetics Clinic
Edmonton Cytogenetics Laboratory
Edmonton Genetics Clinic
Glenrose Rehabilitation Hospital
Northern Alberta Pediatric Cardiology Program
Provincial Newborn Screening Program
Public health units

*Completed by all acute care hospitals in the province.

Discussion

Surveillance and registry systems are apt to become repositories of data that will ultimately be lost if not used actively; however, research is one of the ways that the data can be put to good use. Furthermore, research improves the quality of the data since they are continuously being re-examined and reappraised. A number of projects

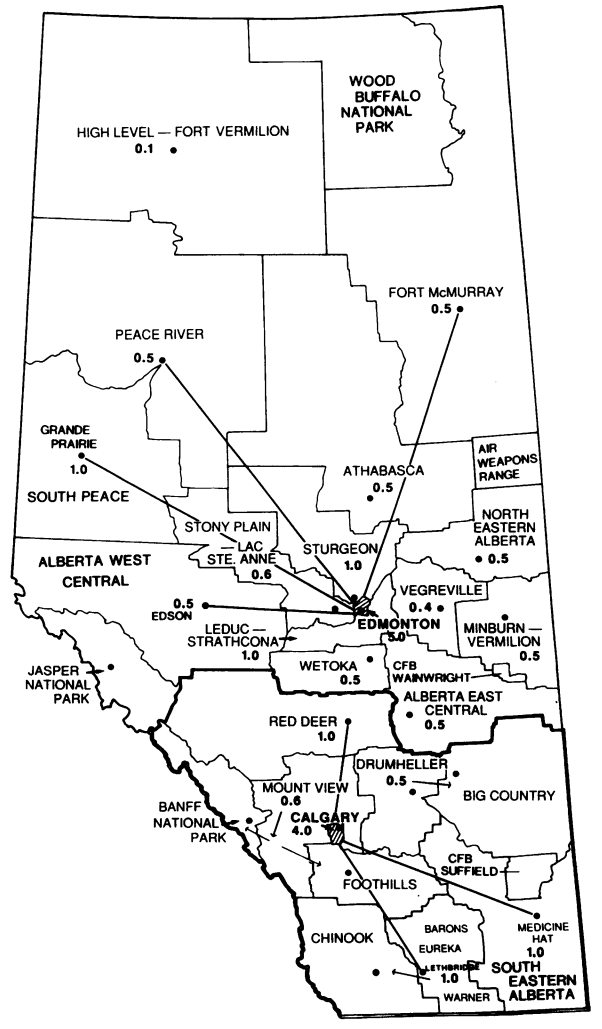

Fig. 1 — Map of Alberta, showing boundaries of health units. CFB = Canadian Forces Base.

have used[5,6] or are using data from the Alberta surveillance system.

The striking difference in the prevalence rates between Edmonton and Calgary may reflect an ascertainment problem, since outpatient sources were added much later for Edmonton than for Calgary and since the 1986 rates were more comparable. However, there may have been a true difference, because the rates of obvious defects, such as anencephaly and spina bifida, differed by 21%; such defects are obvious at birth and thus should be reported immediately and not later on an outpatient basis.

As for the other health units most of the prevalence rates were similar to the provincial average except those for Medicine Hat (Southeastern Alberta Health Unit), where the rates were consistently higher, and for Fort McMurray, where they were much lower. A review of the cases from Medicine Hat did not reveal any major problems, since many of the anomalies were minor and did not appear to be part of any syndrome complex. For example, of the 74 cases of congenital anomalies of the genital organs 41 were of undescended testes and 26 of hypospadias (mostly mild glandular hypospadias). Similarly, among 45 cases of congenital musculoskeletal deformities 11 were of congenital torticollis. Since most teratogens cause a spectrum of anomalies, it is unlikely that extrinsic factors (e.g., drugs and environmental or occupational hazards) caused these isolated cases. Further observation and follow-up are required before one can conclude that the rates were genuinely higher in that health unit.

We were unable to account for the very low rate in Fort McMurray; there were approximately 800 births annually, a rate similar to that for the Alberta West Central and Northeastern Alberta health units, whose rates of anomalies were closer to the provincial average. Perhaps the population in Fort McMurray was different (e.g., young healthy couples who are there because of the oil sands project). The distribution of cases in Fort McMurray was quite even over the 7-year period; this suggests that ascertainment was not a problem early in the study.

We compared the rates of selected defects from the Alberta surveillance system with the rates from three other systems (Table III).[7-9] The rates in Alberta and British Columbia were strikingly similar, with only a few exceptions. The rate of neural tube defects (NTDs) in British Columbia included live births only and thus excluded stillborn infants with anencephaly or spina bifida; it also excluded infants with encephalocele. The rate in Alberta per 1000 live births was 0.47. We presume that the

Table II — Prevalence rates of birth defects reported to Alberta Congenital Anomalies Surveillance System from 1980 to 1986 by health unit

Health unit	No. of births	No. of affected infants*	No. of defects per 1000 total births	95% confidence limits
Alberta East Central	6 437	257	39.93	35.20, 45.12
Alberta West Central	5 795	153	26.40	22.39, 30.93
Athabasca	4 515	141	31.23	26.30, 36.83
Banff	433	18	41.57	24.68, 65.62
Barons–Eureka–Warner	5 374	178	33.12	28.44, 38.36
Big Country	1 530	43	28.10	20.36, 37.88
Calgary Health Services	78 587	3 852	49.02	47.48, 50.59
Chinook	4 472	131	29.29	24.50, 34.76
Drumheller	3 861	155	40.15	34.08, 46.98
Edmonton Board of Health	73 147	2 595	35.48	34.13, 36.87
Foothills	3 778	137	36.26	30.45, 42.86
Fort McMurray and district	5 883	87	14.79	11.51, 18.41
High Level–Fort Vermilion	2 359	66	27.98	21.65, 35.58
Leduc–Strathcona	11 465	377	32.88	29.65, 36.38
Lethbridge	6 350	235	37.01	32.43, 42.05
Minburn–Vermilion	2 751	86	31.26	25.02, 38.63
Mount View	7 965	290	36.41	32.34, 40.85
Northeastern Alberta	5 359	149	27.80	23.53, 32.64
Peace River	5 691	116	20.38	16.85, 24.44
Red Deer	15 075	526	34.89	31.98, 38.01
South Peace	8 775	345	39.32	35.28, 43.69
Southeastern Alberta	9 151	423	46.22	41.93, 50.85
Stony Plain–Lac St. Anne	8 045	245	30.45	26.76, 34.52
Sturgeon	11 361	486	42.78	39.06, 46.76
Vegreville	2 931	65	22.18	17.13, 28.25
Wetoka	4 224	178	42.14	36.19, 48.80
Total	303 478†	11 406†	37.58	36.90, 38.28

*Includes live and still births at 20 weeks' gestation or later.

†Reflects the Department of Vital Statistics total; some normal births and cases were not reported from individual health units because the families may have resided on an Indian reserve or in a national park or because there was no assigned health unit.

rates for total births would be comparable between the two provinces, because that was the case in earlier reports when British Columbia did include stillbirths; the rate was 1.6 per 1000 total births during 1966–81 in British Columbia and 1.62 per 1000 total births during 1970–81 in Alberta.

Although it is unclear why, the rates of orofacial clefting in British Columbia have always been near the upper limit for the white population.[10] The rate among North American Indians in British Columbia is one of the highest in the world; however, these people comprise only 2% of the total provincial population. The rates in British Columbia have remained relatively stable over a 35-year period,[11] and the same stability holds true for Alberta, the rate of 1.62 per 1000 total births during 1980–86 being virtually identical to the rate of 1.67 during 1960–74 (R.B.L.: unpublished data). The rates in Western Australia were similar to those in British Columbia.

The difference in the rates of congenital heart defects between Western Australia and the three other regions is somewhat surprising, because the ascertainment period in Western Australia extends to 6 years of age. However, in Western Australia patent ductus arteriosus is only recorded under special conditions (i.e., if it has been ligated, is associated with other cardiovascular defects or is still present at 6 months of age). The fact that the rate in Alberta was so similar to the rate in British Columbia is striking testimony to the ascertainment process in Alberta, because in British Columbia it extends to all ages. The rates in Alberta, British Columbia and Atlanta were comparable to the rate in California after an extensive 5-year cohort study of approximately 19 000 births.[12] Differences in the rates between these three areas could have been due to ascertainment, definition or coding problems or may have reflected actual differences.

Congenital hip dislocation was much more prevalent in Western Australia than in Alberta and British Columbia, even though the criteria for acceptance of a case in Alberta were similar to those in Western Australia. The various ascertainment periods did not likely contribute to the differences between the regions. In a study in Western Australia two-thirds of the cases were diagnosed in the neonatal period, and of those diagnosed in the postnatal period only 2.7% involved children more than 1 year of age.[13] Perhaps the higher rate in Western Australia was due to the special research project. The investigators found fewer cases in the Aborigine population than in the nonaboriginal population; such a differerence was also observed between the North American Indian and white populations in British Columbia.[14]

The general public, politicians and indeed many health professionals have trouble understanding why the frequency of congenital anomalies is often difficult to determine. Ideally, one would ascertain reports of all cases of congenital anomalies in a given population; unfortunately this never happens, and most jurisdictions have to settle for some compromise. One reason for this is the loss of information by virtue of spontaneous abortions, stillbirths and early neonatal deaths; another is the various definitions of congenital anomalies. The literature has disclosed many different rates of congenital anomalies; these usually reflect variations in methodology rather than in actual frequency.

Knox, Armstrong and Lancashire[15] compared the records of a national system in England and

Table III — Prevalence rates of selected congenital anomalies in four regions from 1980 to 1986

Anomaly	Region; rate (and 95% confidence limits)*			
	Alberta	British Columbia†	Western Australia	Atlanta†
Abdominal wall defects	0.29 (0.23, 0.36)	NR	0.42 (0.32, 0.53)	0.53 (0.44, 0.64)
Anorectal atresia or stenosis	0.33 (0.27, 0.40)	NR	0.50 (0.39, 0.62)	0.38 (0.30, 0.48)
Cleft lip with or without cleft palate	1.00 (0.90, 1.13)	1.26 (1.12, 1.38)	1.25 (1.09, 1.44)	1.05 (0.92, 1.21)
Cleft palate	0.60 (0.52, 0.70)	0.84 (0.69, 0.90)	0.70 (0.58, 0.84)	0.43 (0.35, 0.53)
Cleft palate and lip	1.62 (1.48, 1.77)	2.10 (1.87, 2.20)	1.95 (1.74, 2.18)	1.49 (1.32, 1.67)
Congenital heart defects	12.18 (11.79, 12.58)	13.08 (12.72, 13.44)§	7.00 (6.60, 7.43)	11.25 (10.79, 11.72)
Congenital hip dislocation	2.68 (2.50, 2.87)	3.19 (3.00, 3.41)	6.54 (6.15, 6.95)	NR
Down's syndrome	1.07 (0.95, 1.19)	0.99 (0.88, 1.11)	1.18 (1.02, 1.36)	1.05 (0.91, 1.20)
Esophageal atresia or stenosis	0.24 (0.19, 0.31)	0.27 (0.21, 0.33)	0.28 (0.21, 0.38)	0.27 (0.20, 0.35)
Hydrocephalus	0.61 (0.52, 0.70)	0.49 (0.42, 0.58)	0.62 (0.50, 0.75)	0.92 (0.79, 1.06)
Hypoplastic left heart syndrome	0.28 (0.22, 0.35)	0.27 (0.21, 0.34)	0.19 (0.13, 0.27)	0.34 (0.26, 0.43)
Hypospadias	3.59 (3.29, 3.90)	4.66 (4.33, 5.02)	3.05 (2.79, 3.34)	5.51 (5.07, 5.98)
Limb reductions	0.51 (0.43, 0.59)	0.70 (0.61, 0.80)	0.55 (0.44, 0.68)	0.52 (0.42, 0.63)
Neural tube defects‡	1.01 (0.90, 1.13)	0.73 (0.63, 0.83)	1.90 (1.69, 2.12)	1.27 (1.12, 1.44)
Tetralogy of Fallot	0.19 (0.15, 0.25)	0.41 (0.35, 0.50)	0.31 (0.23, 0.42)	0.36 (0.29, 0.46)

*Alberta and Western Australia rates are per 1000 total births; British Columbia and Atlanta rates are per 1000 live births.
†NR = not reported.
‡Rate does not include cases in stillborn infants or cases of encephalocele.
§Between 1974 and 1983.

Wales with those of a local system in Birmingham, England, which has been in operation for at least 30 years and has multiple sources of ascertainment and a much longer follow-up period. They found that a national congenital anomaly surveillance system is inadequate when it relies on a single reporting source, mainly the birth notice.

For many, if not most, congenital anomalies there is no possibility of total prevention. There appears to be a basic load that every population must carry; this amounts to 3% if reported in the first year of life but 6% to 7% if that cohort is followed up to school entry. Many of the defects are minor or can be corrected (e.g., patent ductus arteriosus). However, since congenital anomalies are the leading cause of death among children 1 to 4 years of age and account for a large proportion of the infant and childhood morbidity rate, it is essential to enumerate the load and have reliable baseline data. There are few such reliable sources with long-term data in North America; probably the best are the British Columbia Health Surveillance Registry[16] and the Congenital Malformation Surveillance of the US Centers for Disease Control, Atlanta.[17] Other countries with centres of excellence include Hungary,[18] Finland,[19] Sweden,[20] Australia (Western Australia)[21] and England (Birmingham).[22]

Surveillance systems have been established to detect new teratogens and would probably have succeeded in detecting thalidomide because it caused severe and distinct limb defects, which are otherwise very rare. However, a new teratogen will more likely be detected by an astute clinician, although one surveillance system in France was responsible for identifying the link between valproic acid and NTDs.[23] Passive surveillance systems (i.e., ones that merely collect data) will not likely detect teratogens, because some type of follow-up or intervention is needed (i.e., an active system) if there are statistically significant deviations from the baseline data.

Surveillance systems with good databases are extremely useful in testing hypotheses regarding teratogens. Investigative studies can be done if a cluster occurs and are much less expensive and more efficient than an ad hoc study of each episode. The public is naturally concerned about the effects of pollution, drugs and increased chemical or radiation exposure, and a good database will alleviate such anxiety. Many stories on birth defects in the media are completely misleading because the reporters and producers fail to realize the complexity of congenital anomalies, particularly with respect to heterogeneity, multiple causes and confidence limits.

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