A B S T R A C T

The Canadian Congenital Anomalies Surveillance System monitors birth defects reported for stillborns, newborns and infants during the first year of life. Data are available through the 1980s and early 1990s for Ontario, Manitoba and Alberta, and since 1984 for an additional four provinces. Fifty-seven routine monitoring categories and 15 summary categories were examined for temporal trends. Comparing the period 1979-1981 with 1991-1993, the reported birth defect case birth prevalence increased by 0.2% and the total birth defects birth prevalences by 2.5%. The birth prevalence of central nervous system defects decreased by 8.2%; the reported birth prevalence increased for congenital heart defects by 41%, urinary defects by 127%, Down syndrome by 13% and other chromosomal defects by 47%. Further investigation of individual defects would be required to evaluate the degree to which changes in reported birth prevalence reflect changes including the availability and use of specific diagnostic procedures. The work highlights the need to expand the surveillance system to include all affected pregnancies where an anomaly has been detected antenatally.

A B R É G É

Le Système canadien de surveillance des anomalies congénitales recense les cas de malformation congénitale déclarés chez les mort-nés, les nouveau-nés et les nourrissons pendant la première année de vie. Des données sont disponibles pour toutes les années 1980 pour Ontario, Manitoba et Alberta et depuis 1984 pour quatre autres provinces. Pour déterminer les tendances temporelles, on a examiné 57 catégories de surveillance systématique et 15 catégories générales. Il ressort de la comparaison des périodes 1979-1981 et 1991-1993 que la prévalence des cas de malformation congénitale déclarés à la naissance a augmenté de 0,2 % et que la prévalence totale des naissances avec malformations congénitales a augmenté de 2,5 %. La prévalence des naissances avec anomalies du système nerveux central a diminué de 8,2 %; la prévalence des naissances déclarées a augmenté pour les malformations cardiaques congénitales (41 %), les malformations du système urinaire (127 %), le syndrome de Down (13 %), et d'autres anomalies chromosomiques (47 %). Il faudra pousser les études sur chaque anomalie afin de déterminer dans quelle mesure les changements enregistrés dans la prévalence des naissances déclarées reflètent des changements tels que la disponibilité et l'utilisation de méthodes diagnostiques précises. Cet ouvrage souligne le besoin d'étendre le champ d'activité des systèmes de surveillance afin d'y inclure toutes les grossesses où une anomalie fut détectée durant la période anténatale.

Temporal Trends in Canadian Birth Defects Birth Prevalences, 1979-1993

Kenneth C. Johnson, PhD,¹ Jocelyn Rouleau²

The Canadian Congenital Anomalies Surveillance System (CCASS)¹⁻³ is a population-based birth defects monitoring system that was initiated in the late 1960s following the rubella epidemic of the 1950s and the tragedy of thalidomide. It was set up to enable the Department of Health and Welfare to monitor birth defect trends and allow it to identify any sudden increases in birth defects, thus alerting officials to possible new teratogenic concerns.

The reporting system underwent several revisions from its inception through the late 1970s. Since the early 1980s, however, defects have been ascertained primarily through automated searching of hospital admission-discharge summary data to determine defects reported in hospital at the time of birth and in the first year of life.⁴ Over the past 15 years CCASS has efficiently assembled, in a manner consistent within provinces, data concerning more than 120,000 infants with birth defects from a pool in excess of three million Canadian births. The current analysis uses that data to examine time trends over the 1980s in reported birth defects birth prevalences in Canada. Few evaluations of birth defect time trends in Canada have been published previously.⁵⁻⁷

It is important to note that data on pregnancy losses prior to 20 weeks gestation and terminations are not included in the database because consistently collected population-based sources of such data have

not been available. Numbers and changes in frequencies of defects and chromosomal abnormalities in these important groups of pregnancies can make interpretation of trends difficult.

METHODS

Data sources for CCASS have varied over time and by province.¹⁻⁴ For the period since 1978, the CCASS database for Ontario has been compiled exclusively from a computer search of hospital admission-discharge summaries (with identifying information removed or consistently scrambled to avoid individual identification) compiled by the Hospital Medical Records Institute (HMRI, responsibilities of HMRI recently assumed by the Canadian Institute for Health Information).¹

The HMRI database includes a record for every separation from a public hospital in Ontario since 1978. Records for all stillbirths and every separation including a diagnostic code for a birth defect for a child under one year of age at the time of discharge are chosen for the birth defect registry. An automated record linkage routine is then used to amalgamate records for the same infant and to resolve individual identifier and birth defect coding conflicts.⁴ The latter can arise when identifying information changes or when diagnoses change in later admissions to hospital.¹ Through the 1980s, other provinces including Saskatchewan, British Columbia, Prince Edward Island and Newfoundland began to have wide HMRI coverage. In Manitoba the Manitoba Hospital Services Plan databases provide data similar to that obtainable from HMRI. Alberta is the only province that has maintained an active reg-

1. Environmental Risk Assessment and Case Surveillance Division, Cancer Bureau, Laboratory Centre for Disease Control, Health Protection Branch, Health Canada
2. Bureau of Reproductive and Child Health, Laboratory Centre for Disease Control

Corresponding author and reprint requests: Kenneth C. Johnson, Cancer Bureau, LCDC Bldg 0601C1, Tunney's Pasture, Ottawa, ON K1A 0L2.

istry, the Alberta Congenital Anomalies Surveillance System, which includes multiple record sources into the 1990s,⁸ and is used for Alberta for CCASS.

Temporal trends are evaluated for the period from 1979 to 1993. The starting year was chosen because by 1979, hospitals in Canada had adopted the Ninth revision of the International Classification of Diseases (ICD-9)⁹ and this revision of the ICD continued to be used through the early 1990s. The year 1993 was the most recent year for which full first-year-of-life follow-up data were available when this analysis was initiated. Consistently collected data are available for the 1980s for three provinces—Ontario, Manitoba and Alberta—and consistently collected data are available since 1984 for an additional four provinces—Newfoundland, Prince Edward Island, Saskatchewan and British Columbia.

CCASS uses 57 specific categories and 15 summary categories of birth defects for routine monitoring. The individual defects and groupings reflect the ICD-9 rubrics. Codes falling within the Congenital Anomalies chapter (ICD-9 codes 740.0-759.9) are included with the exception of undescended testes, skin tags, birth marks and patent *ductus arteriosus*, as these defects are generally trivial, and their degree of reporting varies markedly.

The 72 categories were examined for temporal trends over the 1980s for Ontario, Manitoba and Alberta by comparing the average birth prevalences for 1979-1981 with the average birth prevalence of each defect for 1991-1993. Data for all seven provinces for 1984-1986 were compared with similar data for 1991-1993. Mean annual percentage change was calculated by dividing the total change in birth prevalence by the mean number of years between the two time periods. A chi-square test for trends in proportions¹⁰ was also performed using all individual birth years for the time period being evaluated.

Birth defect birth prevalence is defined as the number of birth defects identified in livebirths and stillbirths at the time of birth or within the first year of life divided by the total number of livebirths and stillbirths. The birth defect case birth prevalence is the number of individual liveborn

and stillborn infants with at least one birth defect divided by the number of individual liveborns and stillborns.

RESULTS

Table I and Figure 1 present the 3-province 12-year temporal trends (1979-1981 to 1991-1993) for the 72 defect categories, ordered essentially as they appear in the Congenital Anomalies chapter of the ICD-9. Table II and Figure 2 present the 7-year trends (1984-1986 to 1991-1993) for the seven provinces combined. Between 1991 and 1993 for the seven provinces combined, there were 44,303 non-trivial birth defects reported among the 33,620 infants with reported defects (1.32 defects/case). This represents a birth defect birth prevalence of 5.6 defects per 100 births and a case rate of 4.2 cases per 100 births.

Comparing 1979-1981 with 1991-1993, reported birth defects case birth prevalences increased by 0.2% and the total birth defect birth prevalence increased by 2.5%. The most important decrease in the 12-year period was observed for birth prevalences of central nervous system defects which, as a group, decreased by 8%. Anencephalus birth prevalence decreased by 53%, spina bifida by 28% and congenital hydrocephalus by 21% (see Figure 3). The large increase in the other specified and unspecified CNS anomaly category was due to an increase in the rates for ICD code 742.4, "other specified anomalies of the brain".

The reported birth prevalence of congenital heart defects as a group increased by 41%, with increases of 20% to 100% in 9 of the 12 specific heart anomaly categories (see Figure 4). The reported prevalence of urinary defects as a group increased by 127%. The reported prevalence of renal agenesis and dysgenesis increased by 24%, cystic kidney prevalence increased 177% and other urinary defects were up 147%. Down syndrome birth prevalence increased 13%, while other chromosomal defects were up 47%. Reporting of trisomies 13 and 18 increased 71% and 61% respectively over the 12 years, but the prevalences are much lower than for Down syndrome (see Figure 5).

The seven-province analyses comparing 1984-1986 birth prevalences with 1991-1993 prevalences reflect a pattern consistent with the 12-year data.

DISCUSSION

Factors affecting the reporting of specific birth defects, including ease, precision and uniformity of diagnosis, classification and coding,¹¹ must always be given serious consideration when attempting to evaluate whether there have been real changes in the occurrence of specific birth defects over time. Readily identifiable defects such as anencephaly and cleft lip are less likely to be affected by changes in reporting than the less obvious or more difficult to diagnose defects. Inter-provincial differences in anomaly rates may result from differences in reporting requirements, coder skill and training, quality assurance mechanisms and person responsible for coding. For example, in Alberta multiple sources for ascertainment are used whereas Ontario and Manitoba rates are based on hospitalization data only. Evaluation of birth defect prevalence when Ontario surveillance was based on multiple sources (physicians' notice of birth, stillbirth certificates, hospitalizations and autopsy reports) suggested that over 90% of reported defects included a hospitalization report (unpublished internal LCDC report).

The availability and extent of use of several prenatal screening techniques, and of selective terminations, are likely to make important contributions to the reported defect birth prevalences, particularly for neural tube defects and Down syndrome. The increase in urinary anomaly reporting is likely the result of the introduction of dramatically improved in utero ultrasound during the 1980s,¹² while increases in cardiac defects may be the result of advances in diagnostic techniques and changes in health care which have increased survival among infants who previously would not have survived to diagnosis.¹¹

Changes in the age distribution of the mothers giving birth will also affect a few birth defect prevalences. An evaluation of 25,859 children with birth defects of unknown aetiology in British Columbia showed maternal-age-specific trends in

TABLE I
Percentage Change in Reported Birth Defect Birth Prevalences* for
Three Canadian Provinces Combined,† 1991-1993 Compared with 1979-1981

Birth Defects	Cases		Rates / 10,000 Births		Mean Annual % Change	12 Year Change	Trend Test‡
	79-81	91-93	79-81	91-93			
Central nervous system	1,712	1,841	31.8	29.2	-0.7%	-8.2%	1
Anencephalus & similar anomalies	240	135	4.5	2.1	-4.4%	-53.3%	1
Spina bifida	470	399	8.7	6.3	-2.3%	-27.6%	1
Encephalocele	79	91	1.5	1.4	-0.6%	-6.7%	
Microcephalus & brain reduction	345	381	6.4	6.0	-0.5%	-6.3%	
Congenital hydrocephalus	486	450	9.0	7.1	-1.8%	-21.1%	1
Other specified & unspecified CNS anom.	289	589	5.4	9.3	6.0%	72.2%	1
Eye anomalies	324	347	6.0	5.5	-0.7%	-8.3%	
Anophthalmos, microphthalmos	71	72	1.3	1.1	-1.3%	-15.4%	
Other eye anomalies	277	293	5.1	4.6	-0.8%	-9.8%	
Ear, face & neck anomalies	816	1,134	15.2	18.0	1.5%	18.4%	1
Anomalies of ear causing impairment	37	55	0.7	0.9	2.4%	28.6%	
Other ear anomalies	553	813	10.3	12.9	2.1%	25.2%	1
Anomalies of face & neck	263	313	4.9	5.0	0.2%	2.0%	
Congenital heart defects	3,016	4,996	56.0	79.1	3.4%	41.3%	1
Common truncus	66	76	1.2	1.2	0.0%	0.0%	
Transposition of great vessels	226	266	4.2	4.2	0.0%	0.0%	
Tetralogy of Fallot	136	268	2.5	4.2	5.7%	68.0%	1
Common ventricle	61	91	1.1	1.4	2.3%	27.3%	
Ventricular septal defect	1,276	1,922	23.7	30.4	2.4%	28.3%	1
Atrial septal defect	625	1,454	11.6	23.0	8.2%	98.3%	1
Endocardial cushion defects	187	179	3.5	2.8	-1.7%	-20.0%	1
Other septal closure defects	28	50	0.5	0.8	5.0%	60.0%	
Heart valve anomalies	424	692	7.9	11.0	3.3%	39.2%	1
Hypoplastic left heart syndrome	115	180	2.1	2.9	3.2%	38.1%	1
Other heart anomalies	1,105	1,577	20.5	25.0	1.8%	22.0%	1
Patent ductus arteriosus	969	1,655	18.0	26.2	3.8%	45.6%	1
Circulatory system anomalies	1,448	1,758	26.9	27.8	0.3%	3.3%	
Coarctation of aorta	211	302	3.9	4.8	1.9%	23.1%	1
Other anomalies of aorta	123	98	2.3	1.6	-2.5%	-30.4%	1
Pulmonary artery anomalies	562	518	10.4	8.2	-1.8%	-21.2%	1
Other circulatory system anomalies	647	930	12.0	14.7	1.9%	22.5%	1
Respiratory system anomalies	542	722	10.1	11.4	1.1%	12.9%	1
Nose anomalies	146	161	2.7	2.6	-0.3%	-3.7%	
Lung agenesis & hypoplasia	295	351	5.5	5.6	0.2%	1.8%	
Other respiratory system anomalies	107	219	2.0	3.5	6.3%	75.0%	1
Cleft lip and/or palate	991	1,073	18.4	17.0	-0.6%	-7.6%	
Cleft palate	392	400	7.3	6.3	-1.1%	-13.7%	
Cleft lip	190	232	3.5	3.7	0.5%	5.7%	
Cleft palate with cleft lip	409	441	7.6	7.0	-0.7%	-7.9%	
Digestive system anomalies	2,531	2,336	47.0	37.0	-1.8%	-21.3%	1
T-E fistula, esophageal atresia & stenosis	145	217	2.7	3.4	2.2%	25.9%	
Other upper alimentary tract anomalies	1,647	1,272	30.6	20.1	-2.9%	-34.3%	1
Intestinal, anorectal atresia & stenosis	255	285	4.7	4.5	-0.4%	-4.3%	
Other digestive system anomalies	576	655	10.7	10.4	-0.2%	-2.8%	
Genital organ anomalies	1,844	2,062	34.3	32.7	-0.4%	-4.7%	1
Hypospadias, epispadias	1,343	1,529	25.0	24.2	-0.3%	-3.2%	
Other genital organ anomalies	517	575	9.6	9.1	-0.4%	-5.2%	
Urinary system anomalies	631	1,680	11.7	26.6	10.6%	127.4%	1
Renal agenesis & dysgenesis	184	265	3.4	4.2	2.0%	23.5%	1
Cystic kidney disease	96	314	1.8	5.0	14.8%	177.8%	1
Other urinary system anomalies	428	1,249	8.0	19.8	12.3%	147.5%	1
Musculoskeletal anomalies	9,121	9,286	169.5	147.1	-1.1%	-13.2%	1
Certain musculoskeletal anomalies	339	354	6.3	5.6	-0.9%	-11.1%	
Congenital dislocation of hip	2,613	2,397	48.6	38.0	-1.8%	-21.8%	1
Clubfoot	2,981	2,824	55.4	44.7	-1.6%	-19.3%	1
Polydactyly, syndactyly	918	1,099	17.1	17.4	0.1%	1.8%	
Limb reduction anomalies	239	293	4.4	4.6	0.4%	4.5%	
Other, unspecified limb anomalies	1,192	1,202	22.1	19.0	-1.2%	-14.0%	1
Anomalies of abdominal wall	210	415	3.9	6.6	5.8%	69.2%	1
Other musculoskeletal anomalies	1,314	1,436	24.4	22.7	-0.6%	-7.0%	
Anomalies of integument	266	513	4.9	8.1	5.4%	65.3%	1
Down syndrome	594	784	11.0	12.4	1.1%	12.7%	
Other chromosomal anomalies	215	373	4.0	5.9	4.0%	47.5%	1
Trisomy 13	36	76	0.7	1.2	6.0%	71.4%	1
Trisomy 18	68	134	1.3	2.1	5.1%	61.5%	1
Autosomal syndromes	55	75	1.0	1.2	1.7%	20.0%	
Sex chromosome conditions	59	95	1.1	1.5	3.0%	36.4%	
Other & unspecified anomalies	748	740	13.9	11.7	-1.3%	-15.8%	
Cases	20,690	24,305	384.4	385.0	0.0%	0.2%	1
All anomalies	27,264	32,796	506.6	519.5	0.2%	2.5%	1

* Reported in the first year of life or at birth.

† The three provinces are Alberta, Manitoba and Ontario, which account for 52% of births in Canada each year 1979-1993 (206,058 births in 1993)

‡ Flag 1 test positive for trend, P-value < 0.01

Figure 1. Percentage change in birth defect prevalences (reported during the first year of life or at birth) for Alberta, Manitoba and Ontario, 1991-1993 compared with 1979-1981.

TABLE II
Percentage Change in Reported Birth Defect Birth Prevalences* for
Seven Canadian Provinces combined,† 1991-1993 Compared with 1984-1986

Birth Defects	Cases		Rates / 10,000 Births		Mean Annual % Change	7 Year Change	Trend Test‡
	84-86	91-93	84-86	91-93			
Central nervous system	2,453	2,347	32.4	29.6	-1.2%	-8.6%	
Anencephalus & similar anomalies	274	172	3.6	2.2	-5.6%	-38.9%	1
Spina bifida	609	544	8.0	6.9	-2.0%	-13.7%	1
Encephalocele	113	108	1.5	1.4	-1.0%	-6.7%	
Microcephalus & brain reduction	535	481	7.1	6.1	-2.0%	-14.1%	
Congenital hydrocephalus	650	540	8.6	6.8	-3.0%	-20.9%	1
Other specified & unspecified CNS anom.	562	757	7.4	9.5	4.1%	28.4%	1
Eye anomalies	544	440	7.2	5.5	-3.4%	-23.6%	1
Anophthalmos, microphthalmos	113	92	1.5	1.2	-2.9%	-20.0%	
Other eye anomalies	456	370	6.0	4.7	-3.1%	-21.7%	1
Ear, face & neck anomalies	1,194	1,428	15.8	18.0	2.0%	13.9%	1
Anomalies of ear causing impairment	64	71	0.8	0.9	1.8%	12.5%	
Other ear anomalies	813	1,009	10.7	12.7	2.7%	18.7%	1
Anomalies of face & neck	363	406	4.8	5.1	0.9%	6.2%	
Congenital heart defects	5,415	6,440	71.5	81.2	1.9%	13.6%	1
Common truncus	86	93	1.1	1.2	1.3%	9.1%	
Transposition of great vessels	357	346	4.7	4.4	-0.9%	-6.4%	
Tetralogy of Fallot	310	349	4.1	4.4	1.0%	7.3%	
Common ventricle	104	115	1.4	1.4	0.0%	0.0%	
Ventricular septal defect	2,272	2,540	30.0	32.0	1.0%	6.7%	
Atrial septal defect	1,108	1,736	14.6	21.9	7.1%	50.0%	1
Endocardial cushion defects	349	247	4.6	3.1	-4.7%	-32.6%	1
Other septal closure defects	73	69	1.0	0.9	-1.4%	-10.0%	
Heart valve anomalies	748	834	9.9	10.5	0.9%	6.1%	1
Hypoplastic left heart syndrome	200	212	2.6	2.7	0.5%	3.8%	
Other heart anomalies	1,764	2,004	23.3	25.3	1.2%	8.6%	
Patent ductus arteriosus	1,756	2,032	23.2	25.6	1.5%	10.3%	1
Circulatory system anomalies	2,323	2,285	30.7	28.8	-0.9%	-6.2%	
Coarctation of aorta	370	384	4.9	4.8	-0.3%	-2.0%	
Other anomalies of aorta	187	129	2.5	1.6	-5.1%	-36.0%	1
Pulmonary artery anomalies	627	670	8.3	8.4	0.2%	1.2%	
Other circulatory system anomalies	1,304	1,218	17.2	15.4	-1.5%	-10.5%	1
Respiratory system anomalies	982	928	13.0	11.7	-1.4%	-10.0%	
Nose anomalies	234	217	3.1	2.7	-1.8%	-12.9%	
Lung agenesis & hypoplasia	606	471	8.0	5.9	-3.7%	-26.2%	1
Other respiratory system anomalies	159	251	2.1	3.2	7.5%	52.4%	1
Cleft lip and/or palate	1,459	1,507	19.3	19.0	-0.2%	-1.6%	
Cleft palate	507	535	6.7	6.7	0.0%	0.0%	
Cleft lip	299	344	3.9	4.3	1.5%	10.3%	
Cleft palate with cleft lip	653	628	8.6	7.9	-1.2%	-8.1%	
Digestive system anomalies	3,709	3,121	48.9	39.3	-2.8%	-19.6%	1
I-E fistula, esophageal atresia & stenosis	292	266	3.9	3.4	-1.8%	-12.8%	
Other upper alimentary tract anomalies	2,265	1,692	29.9	21.3	-4.1%	-28.8%	1
Intestinal, anorectal atresia & stenosis	374	390	4.9	4.9	0.0%	0.0%	
Other digestive system anomalies	924	885	12.2	11.2	-1.2%	-8.2%	
Genital organ anomalies	2,599	2,697	34.3	34.0	-0.1%	-0.9%	
Hypospadias, epispadias	1,904	1,964	25.1	24.8	-0.2%	-1.2%	
Other genital organ anomalies	736	785	9.7	9.9	0.3%	2.1%	
Urinary system anomalies	1,439	2,341	19.0	29.5	7.9%	55.3%	1
Renal agenesis & dysgenesis	349	381	4.6	4.8	0.6%	4.3%	
Cystic kidney disease	273	389	3.6	4.9	5.2%	36.1%	1
Other urinary system anomalies	994	1,772	13.1	22.3	10.0%	70.2%	1
Musculoskeletal anomalies	13,948	13,579	184.1	171.2	-1.0%	-7.0%	1
Certain musculoskeletal anomalies	518	474	6.8	6.0	-1.7%	-11.8%	
Congenital dislocation of hip	4,754	4,296	62.7	54.2	-1.9%	-13.6%	1
Clubfoot	4,130	3,742	54.5	47.2	-1.9%	-13.4%	1
Polydactyly, syndactyly	1,234	1,410	16.3	17.8	1.3%	9.2%	
Limb reduction anomalies	377	375	5.0	4.7	-0.9%	-6.0%	
Other, unspecified limb anomalies	1,753	1,784	23.1	22.5	-0.4%	-2.6%	
Anomalies of abdominal wall	295	479	3.9	6.0	7.7%	53.8%	1
Other musculoskeletal anomalies	2,123	2,003	28.0	25.2	-1.4%	-10.0%	1
Anomalies of integument	417	659	5.5	8.3	7.3%	50.9%	1
Down syndrome	947	1,049	12.5	13.2	0.8%	5.6%	
Other chromosomal anomalies	465	506	6.1	6.4	0.7%	4.9%	
Trisomy 13	80	93	1.1	1.2	1.3%	9.1%	
Trisomy 18	156	172	2.1	2.2	0.7%	4.8%	
Autosomal syndromes	111	112	1.5	1.4	-1.0%	-6.7%	
Sex chromosome conditions	128	139	1.7	1.8	0.8%	5.9%	
Other & unspecified anomalies	1,101	1,050	14.5	13.2	-1.3%	-9.0%	
Cases	31,832	33,620	420.1	423.8	0.1%	0.9%	
All anomalies	43,104	44,303	568.8	558.4	-0.3%	-1.8%	

* Reported in the first year of life or at birth

† The seven provinces are British Columbia, Alberta, Saskatchewan, Manitoba, Ontario, Prince Edward Island and Newfoundland, which account for 70% of births in Canada each year 1979-1993 (273,053 births in 1993)

‡ Flag 1 test positive for trend, P-value < 0.01

Figure 2. Percentage change in birth defect prevalences (reported in the first year of life or at birth) for British Columbia, Alberta, Saskatchewan, Manitoba, Ontario, Prince Edward Island and Newfoundland combined, 1991-1993 compared with 1984-1986.

only 3 of 43 birth defect categories studied.¹³ Patent *ductus arteriosus* and hypertrophic pyloric stenosis showed decreasing linear trends with increasing maternal age, while congenital hip dislocations and hip clicks showed a bell-shaped curve. There is a well-known relationship of maternal age to Down syndrome rates. Although as a rule, pregnant women 35 years and older

are tested for Down syndrome during pregnancy, increasing average maternal age has resulted in larger percentages of delivering women between 30 and 34, an age range when the risk of a Down Syndrome child is still small and much lower than at older maternal ages, but 80% higher than for women in their twenties.¹⁴ In Ontario between 1979 and 1989, the percentage of

women giving birth between ages 30 and 34 had increased over 40% (from 18 to 26% of all Ontario births), between 35 and 39 it had doubled and in the 40 to 44 year age range it was up by 45%.^{15,16}

A birth defect trend study done in the United States,¹¹ using data from a national program that monitors defects in approximately 15% of the U.S. population, looked at a more restricted set of defects but found a number of similar trends. Because the U.S. system depends on diagnosis of newborns only, the Canadian data with a full-year follow-up is not necessarily comparable for some defects. Trisomy 13 and 18 show the same dramatic increases in reporting in both countries. Anencephalus and spina bifida show decreasing prevalences in both systems, with the anencephalus rate dropping in both countries at an average of approximately 5% per year. In the U.S. program, all reported cardiovascular defects increased, the majority increasing 10% to 20% per year. In the Canadian data, the increases were of a slightly lower magnitude. As the initial Canadian rates were often several times as high as the U.S. rates and still considerably higher in the late 1980s, it is extremely likely that increased reporting in the newborn period (as opposed to increased birth prevalence) accounts for the dramatic increases in the U.S.

Initiated in response to the thalidomide tragedy and the rubella epidemic, congenital anomalies surveillance systems now exist in many parts of the world. Since their inception, these systems have contributed to the detection of other birth defects resulting from teratogenic compounds.⁷ Today, prenatal diagnostic techniques such as amniocentesis, chorionic villus sampling, and ultrasonography have become increasingly accurate in the diagnosis of birth defects. While detection rates vary depending on the anomaly, a growing list of more than 300 birth defects can be diagnosed prenatally.¹⁷ For example, the prenatal detection rate of Down syndrome is between 45 and 75%, while nearly 100% of anencephaly can be detected prenatally with ultrasonography.¹⁸ The Society of Obstetricians and Gynaecologists of Canada currently recom-

Figure 5. Trends in chromosomal anomalies reported birth prevalences, Ontario, Manitoba and Alberta, 1979-1993.

mends a full screening ultrasound be offered to all women at 16 to 20 weeks gestation, considered the optimal time for evaluation of dating, biometry, as well as the detection of malformations.¹⁹

These increased detection capabilities have had an impact on the numbers of anomalies observed at birth, and the implications for surveillance systems are important to consider. It is possible that anomalies that may have gone undetected for years or perhaps for the whole life span of the individual can now be detected antenatally and recorded at the time of birth. In other cases, prenatal detection of an anomaly or anomalies may result in termination of the affected pregnancy, meaning that these anomalies will not be recorded at birth.

It is hoped that in the near future, congenital anomalies surveillance systems will have the capacity to include counts of affected pregnancies where an anomaly has been detected antenatally. This will enable these systems to better serve the important purpose of prevention of birth defects

through the timely detection of outbreaks and epidemics due to environmental hazards and new substances.

ACKNOWLEDGEMENTS

We thank the Hospital Medical Records Institute (HMRI) (now the Canadian Institute for Health Information), the Alberta Congenital Anomalies Surveillance System and the Manitoba Health Department for providing birth defect data. Thanks to Dr. P.T. Mohide for comments on an early draft of the manuscript and to Linda Turner, Ph.D. for input on prenatal diagnosis.

REFERENCES

1. Sherman GJ. The Canadian Congenital Anomalies Surveillance System: A brief description. *Chron Dis Can* 1982;2:48-50.
2. Sherman GJ. The Canadian Congenital Anomalies Surveillance System (CCASS). Four years later. *Chron Dis Can* 1986;6:76-77.
3. Sherman GJ. Canadian Congenital Anomalies Surveillance System (CCASS). *Chron Dis Can* 1988;8:58-60.
4. Rouleau J, Arbuckle TE, Johnson KC, Sherman GJ. Description and limitations of the Canadian

5. Lowry RB, Trimble BK. Incidence rates for cleft lip and palate in British Columbia 1952-71 for North American Indian, Japanese, Chinese and total populations: Secular trends over twenty years. *Teratology* 1977;16:277-83.
6. Lowry RB, Thunem NY, Uh SH. Birth prevalence of cleft lip and palate in British Columbia between 1952 and 1986: Stability of rates. *Can Med Assoc J* 1989;140:1167-70.
7. Congenital Malformations Worldwide: A Report from the International Clearinghouse for Birth Defects Monitoring Systems; A Non-Governmental Organization of the World Health Organization. Amsterdam: Elsevier Science Publishers Co. Inc. 1991.
8. Lowry RB, Thunem NY, Anderson-Redick S. Alberta Congenital Anomalies Surveillance System; *CMAJ* 1989;141(December 1):1158-59.
9. Ninth Revision Conference, 1975, *Manual of the International Statistical Classification of Diseases, Injuries, and Causes of Death*, Vol.1. Geneva: World Health Organization, 1977.
10. Armitage PA, Berry G. *Statistical Methods in Medical Research*. Oxford: Blackwell Scientific Publications, 1994.
11. Edmunds LD, James LM. Temporal trends in the prevalence of congenital malformations at birth based on the Birth Defects Monitoring Program, United States 1979-1988. *MMWR* 1990;39:19-23.
12. Goncalves LF, Jeanty P, Piper JM. The accuracy of prenatal ultrasonography in detecting congenital anomalies. *Am J Obstet Gynecol* 1994;171:1606-12.
13. Baird PA, Sadovnick AD, Yee IML. Maternal age and birth defects: A population study. *Lancet* 1991;337:527-30.
14. Kline J, Stein Z, Susser M. *Conception to Birth: Epidemiology of Prenatal Development*. Oxford: Oxford University Press, 1989.
15. Vital Statistics, Volume 1, Births and Deaths 1979. Ottawa: Statistics Canada, 1981. Catalogue 84-204 (Annual).
16. Health Report; Supplement No. 14: Births 1989. Ottawa: Statistics Canada, 1991 Volume 3 No 1.
17. Demianczuk NN. Ultrasound diagnosis of congenital anomalies. Presented at the Society of Obstetricians and Gynaecologists of Canada 52nd Annual Clinical Meeting, Quebec City, June 1996.
18. McKay IF, Fraser FC. The history and evolution of prenatal diagnosis. In: *Prenatal Diagnosis: Background and Impact on Individuals*, Royal Commission on New Reproductive Technologies, Volume 12 of the Research Studies. Ottawa: Minister of Supply and Services Canada, 1993.
19. Society of Obstetricians and Gynaecologists of Canada. SOGC Clinical Practice Guidelines for Obstetrics, guidelines for the performance of ultrasound in obstetrics and gynaecology. Policy Statement, No.8, 1994.

Received: June 16, 1995

Accepted: January 12, 1997