Alberta Congenital Anomalies Surveillance System 1980 - 1998

REPORT

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HEALTH AND WELLNESS

December 2001

Alberta Congenital Anomalies Surveillance System

Fifth Report, 1980-1998

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We are housed in space associated with the Department of Medical Genetics at the Alberta Children's Hospital, Calgary Health Region (CHR). We would like to thank the Department and the CHR for their continuing support.

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EXECUTIVE SUMMARY

1. This is the fifth in a series of reports detailing the prevalence of congenital anomalies in Alberta for the calendar years 1996, 1997 and 1998 although aggregate data from 1980 onwards is also included. The main emphasis is on items in Section XIV (congenital anomalies) of the International Classification of Diseases, 9th Edition (ICD-9) although some selected items from other sections have also been included.

2. The inclusion of data relating to fetal anomalies which began in 1997 is a new item in the report. The intent of adding this information is to document the frequency and distribution of congenital anomalies diagnosed in fetuses less than 20 weeks gestation in order to give us a better estimate of the true incidence of congenital anomalies such as neural tube defects (NTD's) or chromosome abnormalities.

3. The overall frequency of most congenital anomalies is relatively unchanged with the exception of anencephaly, which continues to decline probably as a result of termination of pregnancy. The same decline is not seen for other NTD's such as spina bifida. Periconceptional intake of folic acid can reduce the frequency of NTD's however Alberta does not have a major campaign to promote the use of folic acid by women who are at risk of becoming pregnant.

4. Our format has changed from previous reports as we have omitted the histograms which were essentially a duplication of the line graphs. Also, we have added textual material contiguous with the statistics of some of the major sentinel anomalies. Our rates are comparable to many other international registries and surveillance systems, thus corroborating the quality of our data. (see Appendix 7)

5. The percentage of live births to women aged 35 years or over continues to rise and is now approaching 14 per cent. Twenty years ago this was about 4 per cent. Advanced maternal age is associated with an increase of chromosome anomalies and this is reflected in a significant increase in the frequency of Down Syndrome (see section 2.1.4 and Appendix 8).

6. The only other congenital anomaly that appeared to have increased is limb reductions. Unfortunately, this is a very heterogeneous category and can range from absence of part of a finger to a missing arm or leg. Fluctuations in the frequency of limb reductions have been

seen in this province previously but investigations have never been able to uncover any particular causes or factors. The same situation pertains in many other registries and surveillance systems, i.e., fluctuating frequencies with no apparent known cause.

7. In contrast to advanced maternal age for Down Syndrome and for some other congenital anomalies the occurrence of gastroschisis is more frequent in younger mothers, i.e., less than 20 years of age. Again, this finding has been noted in several other jurisdictions.

8. Coding of anomalies is done with the British Paediatric Adaptation of the ICD-9 system. Beginning January 1, 2001, ACASS will be changing to ICD-10 coding of anomalies and more particularly to the Royal College of Child Health and Paediatrics adaptation of ICD-10.

9. Coding of minor malformations is an ongoing problem. In many registries and surveillance systems, including ours, there are exclusion lists which outline defects which are not generally acceptable, e.g., patent ductus arteriosus or undescended testes in an infant less than 37 weeks gestation. These lists also include minor anomalies which are rejected if they are the only feature of a case but are accepted and coded if there is also a major congenital anomaly present. Minor anomalies are useful in helping to make a syndrome diagnosis but this requires the active participation of a clinician knowledgeable in syndromes and also the ability to gain access to the case in question without violating any confidentiality issues.

10. Of note, the Regional Health Authority (RHA) boundaries changed in 1998 most dramatically for Regions 9 (Crossroads) and 10 (Capital). The case and anomaly rates for the RHAs for 1998 were calculated based on the new boundaries (numerator and denominator). Therefore it will be difficult to compare rates from pre 1998 to the years that follow especially with respect to Regions 9 and 10.

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1.0 INTRODUCTION

1.1 History

The forerunner of the Alberta Congenital Anomalies Surveillance System (ACASS) was established in 1966 as a Registry for Handicapped Children, along with similar systems in British Columbia, Manitoba and New Brunswick, as part of the Canadian Congenital Anomalies Surveillance System (CCASS). The system was formed in response to the birth of many infants with severe limb defects following mothers' ingestion of thalidomide during pregnancy (1958-1962). The Division of Vital Statistics in the Department of Social Services and Community Health administered the Registry and the first Director, Dr. Charlotte Dafoe, provided medical input. There was essentially no activity for two years after Dr. Dafoe's retirement in 1977. In 1979 the Department decided to downsize the registry into a surveillance system for congenital anomalies. Dr. Tom Paton, who had joined Social Services and Community Health provided direction and medical advice from 1979-1981. Dr. R.B. Lowry, the current medical consultant, was appointed in 1982. The Division of Vital Statistics remained responsible for the initial collection of data however in 1986 financial constraints in that Division put the continued existence of the Surveillance System in jeopardy since personnel could no longer be spared to code the data. This task was then entrusted to the staff of the Southern Alberta Hereditary Diseases Programme (HDP). The System was then partially funded by Vital Statistics with financial assistance from the HDP and two voluntary agencies, the MSI Foundation and the Alberta Children's Hospital Foundation. In 1989, when Vital Statistics could no longer fund the programme, the Alberta Children's Hospital Foundation assumed complete funding of ACASS. In 1996, the Foundation decided that it was no longer able to support the System and it was at this time that Alberta Health and Wellness, Health Surveillance Branch, assumed financial responsibility. We still work closely with Vital Statistics, relying on them to provide us with notifications of birth, death and stillbirth.

1.2 Purpose of a Surveillance System

Public health surveillance is the ongoing, systematic collection, analysis, and interpretation of data (e.g. regarding agent/hazard, risk factor, exposure, health event) essential to the planning, implementation, and evaluation of public health practice, closely integrated with the timely dissemination of these data to those responsible for prevention and control.

The purpose of surveillance for congenital anomalies (CAs) is to:

- provide reliable and valid baseline data of congenital anomalies in Alberta;
- investigate any significant temporal or geographic changes in the frequency of congenital anomalies with a view to identifying environmental, and therefore, possible preventable causes;
- measure trends;
- assess the effectiveness of prevention e.g. folic acid or antenatal screening;
- assist with health related program planning and development through the provision of data.

As well, it is important to look at patterns or associations of malformations to determine whether they belong to an existing or new syndrome complex.

A principle feature of a surveillance system is timeliness, however data collection and analysis should not be accomplished at the expense of accurate diagnosis.

1.3 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 7th week of gestation) and others in the fetal period (8th week to term). The term "anomaly" covers all the major classes of abnormalities of development, of which there are four major categories as follows:

1. 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).
2. 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).
3. 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).
4. Dysplasia – the abnormal organization of cells into tissues and its morphologic result (e.g. Marfan Syndrome, osteogenesis imperfecta).

1.4 Ascertainment

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

As several malformations may occur in the same infant, it is desirable 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).

ACASS 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 both ease and completeness of ascertainment as well as the accuracy of the diagnostic data.

ACASS screens many important Alberta Health and Wellness and Alberta Vital Statistics documents for the presence of a congenital anomaly. These documents include:

- Notice of a Live birth or a Stillbirth and Newborn Record often referred to as the Physician's Notice of Birth (PNOB)
- Medical Certificate of Stillbirth
- Medical Certificate of Death

All acute care hospitals in the province notify Vital Statistics of the live birth, stillbirth, admission or hospital death of an infant under one year of age who has a congenital anomaly. A notification form called the Congenital Anomaly(ies) Reporting Form (CARF) is completed by the hospital health records personnel following the birth or an admission of an affected child. This form serves as the single most important source of case ascertainment.

Since many children with congenital anomalies are not admitted to hospital, it is very important to obtain out-patient information such as from the Calgary Department of Medical Genetics.

Ascertainment at a continued high level requires each hospital record department and each health care provider to co-operate with the system by notifying us as promptly as possible. We are fortunate in having such co-operative agencies and personnel.

1.5 Quality Control Measures

When a copy of a reporting document reaches the ACASS office in Calgary, it is scanned for content by the Research Assistant and Manager. If the information is unclear, such as a vague or queried diagnosis, the Manager, on behalf of the Medical Consultant, writes to the physician

responsible for the case seeking clarification. A postage paid, addressed envelope is included with the letter and the physician is asked to respond at the bottom of the letter thus making the mechanics of replying easy. The response from physicians has been very satisfactory (greater than 90 per cent) and usually this is sufficient to make a decision whether to accept or reject an anomaly or case. Any questionable diagnosis that is not confirmed is rejected from the database. Some cases are also rejected if they contain diagnoses that do not belong in a congenital anomaly system or are part of a normal developmental process such as a patent ductus arteriosus or undescended testes in a premature infant. Any reports requiring a medical decision are referred to the Medical Consultant. Policy decisions with respect to acceptance or rejection of a case and coding are referred to the ACASS Advisory Committee which is comprised of a paediatric cardiologist, neonatologist/epidemiologist, paediatric pathologist, medical geneticist (medical consultant) with occasional input from a paediatric neurologist, paediatric nephrologist and a paediatric orthopaedic surgeon.

1.6 Coding

Coding is done at the Calgary office using the British Paediatric Association (BPA) adaptation of the International Classification of Diseases, ninth edition (ICD-9). We are currently also using the Royal College of Paediatrics and Child Health (RCPCH) adaptation of the soon to be introduced International Classification of Diseases, tenth edition (ICD-10). Difficult cases are referred to the Medical Consultant (Medical Geneticist). In the past we were able to code only 6 anomalies but since 1997 we have been coding all eligible anomalies reported to us.

Coding with respect to neural tube defects is done hierarchically in that an infant born with anencephaly and spina bifida will be coded as anencephaly only. This policy is consistent with other international organizations.

1.7 Confidentiality and Release of Data

Notifications of Congenital Anomalies are sent to Health Surveillance, Alberta Health and Wellness and from there to the ACASS office in Calgary where the database is maintained. The notifications are handled by the Manager, Research Assistant, Secretary and Medical Consultant only. The data are treated in a completely confidential manner and the notifications are kept in locked files in a locked room. The database is secured by limited access and is password protected. Identifying data is not disclosed. Should further clarification become necessary, communication is with the attending physician or physician responsible for ongoing

care. Direct contact is never made with the family. When data are requested, they are released in statistical form with no personal identifiers.

1.8 Methodology and Limitations

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

$$\text{Anomaly (Defect) Rate} = \frac{\text{Number of a particular congenital anomaly among live births and stillbirths}}{\text{Total number of live births and stillbirths}} \times 1000$$

$$\text{Case Rate} = \frac{\text{Number of individual infants (live - or stillborn) with} \geq 1 \text{ congenital anomaly}}{\text{Total number of live births and stillbirths}} \times 1000$$

Confidence intervals (approximate 95 per cent) 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 per cent probability that the true population rate will fall.

One of the major limitations of the surveillance system is that on its own, the information provided to us does not allow studies to determine aetiology. If increasing trends indicate there is a potentially serious problem, then separate investigative studies need to be done. However, it is possible to conduct linkage studies with other data sources to explore potential causes of specific birth defects.

2.0 DATA

2.1 Trends

The following table and graphs of selected sentinel anomalies indicate the trends in congenital anomaly rates in Alberta from 1980 through 1998. Sentinel anomalies are those which the International Clearinghouse of Birth Defects Monitoring Systems watches worldwide, the rationale being they are quite easily identified and therefore will be accurately reported.

Table 1. Chi Squared Linear Trend Analysis and p-values for Selected Anomalies 1980-1998 inclusive (Live Births and Stillbirths).

Anomaly	Trend Direction	Chi Squared Analysis (χ^2_{LT})	p-value
Neural Tube Defects	Decreasing	10.9	0.0009
Anencephaly	Decreasing	16.7	0.0000
Spina Bifida	No significant change	1.5	n.s.
Hydrocephalus	Decreasing	10.3	0.0014
Cleft Lip ± Cleft Palate	No significant change	1.1	n.s.
Cleft Palate	Increasing	4.3	0.0386
Esophageal Atresia/Stenosis	No significant change	0.7	n.s.
Anorectal Atresia/Stenosis	No significant change	1.8	n.s.
Hypospadias and Epispadias *	No significant change	0.02	n.s.
Limb Reductions	Increasing	5.7	0.0170
Abdominal Wall Defects	No significant change	1.3	n.s.
Down Syndrome	Increasing	6.7	0.0099
Renal Agenesis	No significant change	2.2	n.s.
Hypoplastic Left Heart Syndrome	No significant change	0.9	n.s.

* Hypospadias/Epispadias calculated for male live births only

2.1.1 Neural Tube Defects

The data indicate a very slight decline in the overall frequency of neural tube defects (NTD's), however this is virtually all due to the decline in the frequency of anencephaly with essentially no change for spina bifida over the last 18 years. Studies in other parts of the world have indicated declining frequencies in part due to termination of pregnancies and in part due to very active promotion and preventive effects following the periconceptional use of folic acid (taken prior to conception and throughout the first trimester). The United States began fortifying flour and cereal/grain products in 1996 and Canada followed suit in 1998. The amount of

fortification (140mcg/100 gm) is considered to be insufficient to give adequate protective effect for pregnant women therefore they should take a folic acid supplement.

Poor nutrition has been associated with NTD's and is probably primarily due to low folate levels as it is now definitely established that the periconceptional use of folic acid reduces the first occurrence and the recurrence of NTD's. The recommended dose is 0.4 mg daily for a woman with a negative family history for NTD's and 4 mg daily to prevent recurrence (Pharmacies stock 1 mg and 5 mg tablets and there is no danger in using these dosages). Other risk factors include poorly controlled diabetes mellitus and anticonvulsant medication such as Valproic Acid and Phenytoin. There is a weak association with advanced maternal age with some studies showing a positive association and others not. Some newer risk factors include the presence of pre-pregnancy maternal obesity (body mass index ≥ 29 kg/m^2), maternal smoking and possibly hyperthermia. The presence of homozygosity for a specific genetic mutation for methylene-tetrahydrofolate reductase (MTHFR) may increase the risk but routine testing for this genetic polymorphism is not yet available. The presence of a previously affected child or if one parent is affected increases the risk from 1/800 (population risk) to approximately 2 per cent. Accurate prenatal screening and/or testing is available, e.g., triple screen (afp, estriol and hcg) at 15 - 16½ weeks gestation plus a detailed ultrasound scan at 18 weeks. An amniocentesis for estimation of amniotic fluid alpha fetoprotein (AFP) is rarely required today.

Figure 1. Neural tube defect rate per 1000 total births, 1980 – 1998.

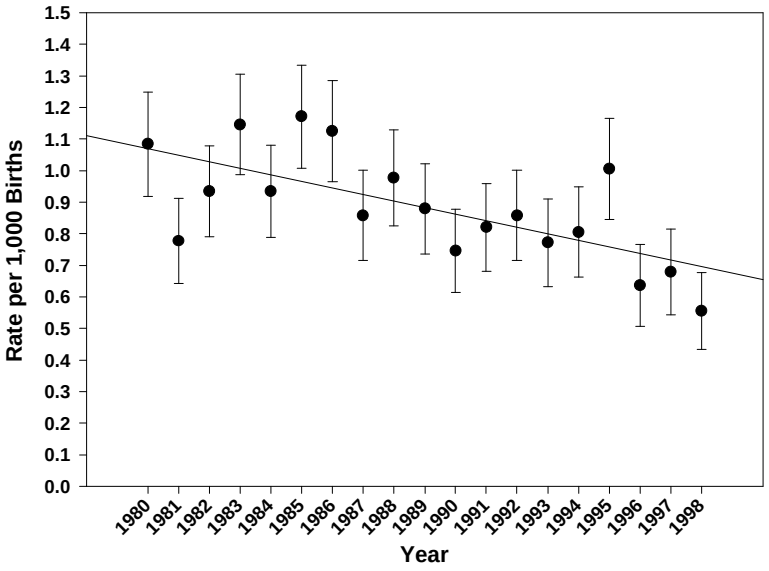


Figure 2. Anecephaly rate per 100 total births, 1980 – 1998.

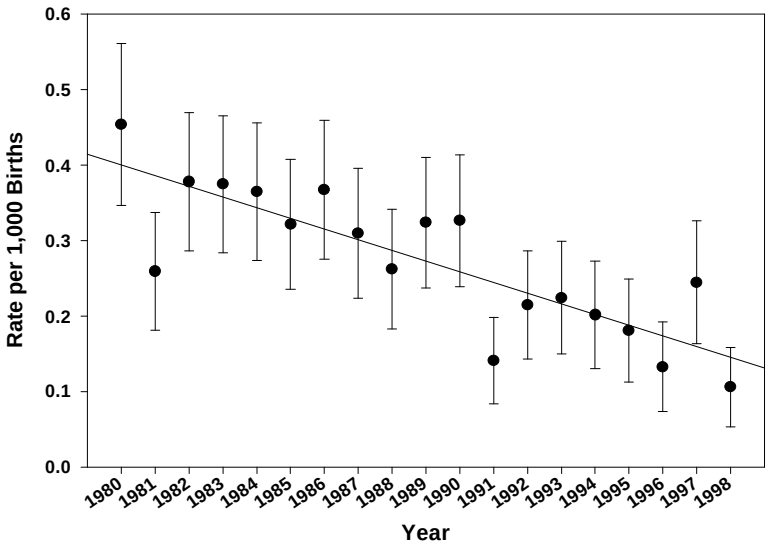
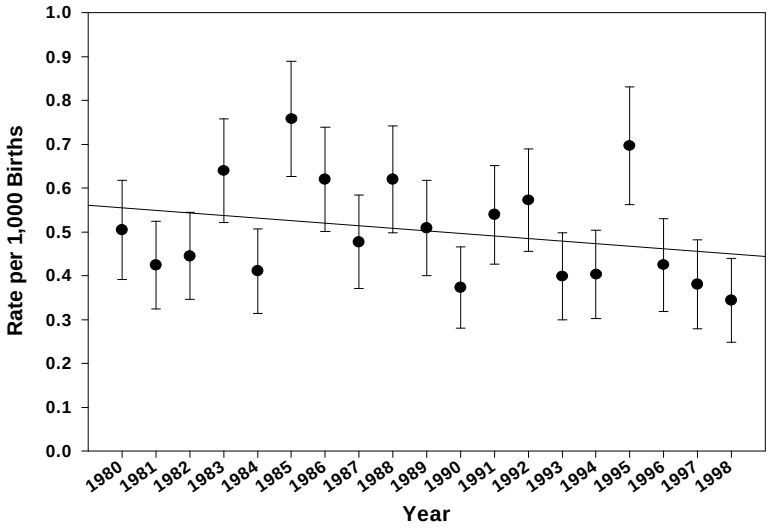


Figure 3. Spina bifida rate per 1000 total births, 1980 – 1998.



2.1.2 Cleft Lip and Palate and Cleft Palate

The birth prevalence of cleft lip with or without cleft palate (CL ± CP) has essentially remained stable over the last 18 years. This was also noted in studies from British Columbia over 35 years suggesting that environmental factors play less of a role in causation than genetic ones. Despite a large number of genetic studies showing either association or linkage studies of orofacial clefts with a large number of chromosomal sites there has been no major breakthrough with respect to aetiology. Two areas show susceptibility loci for non syndromic CL ± CP, one being the short arm of chromosome 6 and the other the short arm of chromosome 2 (transforming growth factor alpha gene). These linkage and association studies have not yet been able to identify random families as being high risk or not. About 70 per cent of CL ± CP are non syndromic and of the remainder there is a mixture of chromosomal disorders, a large number of Mendelian syndromes (more than 300), a few teratogens plus some uncategorized syndromes. Some known teratogens include Phenytoin, valproic acid, thalidomide, alcohol and retinoic acid (Accutane™).

The role of folic acid in preventing recurrence in couples with a positive family history was first shown nearly twenty years ago but involved a very high dose of folic acid, namely 10 mg daily. Since then other studies have shown a reduction in first occurrence with the use of multivitamins including folic acid but it is not clear whether it is the folic acid or some of the other vitamins or trace elements that are present that are responsible for the reduction. It would seem prudent to utilize a periconceptional routine comparable to that employed for NTD's but

making sure that it is not just folic acid but folic acid plus a multivitamin compound. Excessive maternal alcohol is of course known to cause the Fetal Alcohol Syndrome (FAS) and fetal alcohol effect and orofacial clefting occasionally occurs in FAS. An increasing number of studies are confirming an increase risk of orofacial clefting with maternal smoking. Some studies show a positive association with CL ± CP or Cleft Palate (CP) when they occur as isolated anomalies but no association if the orofacial clefting is associated with other malformations. Since both CL ± CP and isolated CP can be part of a number of syndromes it is most important that if other anomalies are present that the child be referred to appropriate pediatric and/or genetic resources to establish if a syndrome is present as this could very well change the management including the recurrence risk.

In the absence of gene markers which are suitable for routine clinical use, we are forced to rely on empiric recurrence risk figures for non syndromic CL ± CP and isolated CP. Thus if two unaffected parents with one CL ± CP affected child or one affected parent with no affected children ask about recurrence risks, we generally quote 2 – 5 per cent depending on the severity of the cleft. Comparable risks for isolated cleft palate are 1 – 2 per cent. It is important to take a detailed family history and to be sure the affected person does not have other anomalies as these would suggest the possibility of a syndrome (genetic or teratogenic).

With respect to prenatally identified facial clefts (by ultrasound), associated anomalies occur in more than 50 per cent of fetuses, depending on the type of cleft observed. The risk for fetal aneuploidy especially Trisomy 13 is high when facial clefts are identified.

Figure 4. Cleft Lip +/- cleft palate rate per 1000 total births, 1980 – 1998.

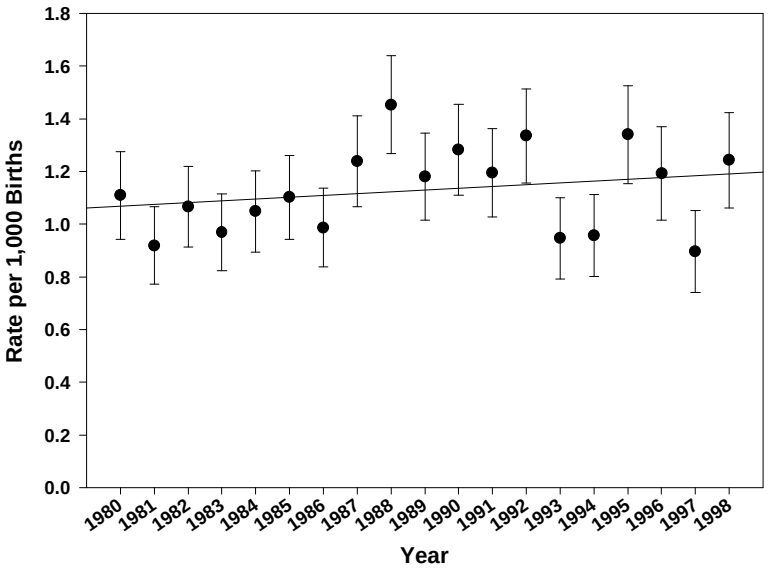
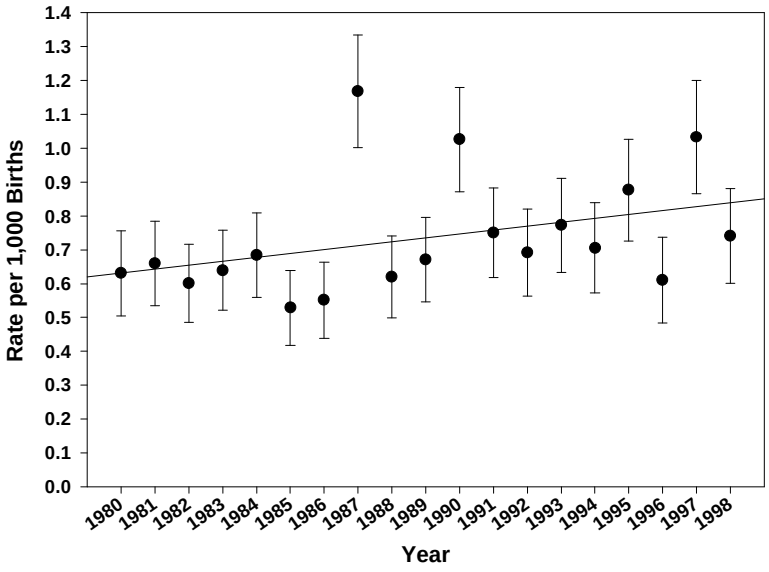


Figure 5. Cleft palate rate per 1000 total births, 1980 – 1998.



2.1.3 Abdominal Wall Defects

This includes both omphalocele and gastroschisis. It is vitally important to differentiate between them as omphalocele is more likely to be found in association with other anomalies or syndrome complexes. The overall frequency of abdominal wall defects shows no significant upward or downward trend, however, gastroschisis is one of the few defects which is associated with an increased frequency in younger mothers (see Table 2). These findings have also been reported in several other jurisdictions.

Table 2. Gastroschisis by Maternal Age, 1989-1998 inclusive.

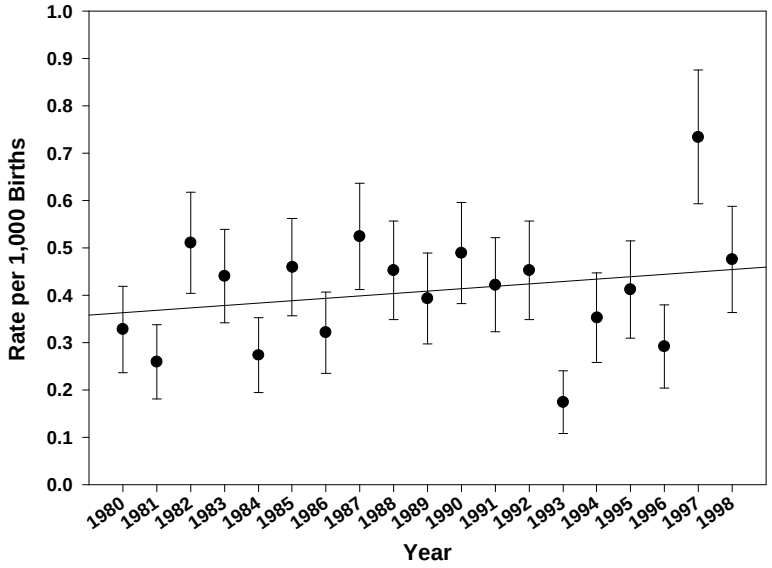
Maternal Age (years)	Number	Rate per 1000 total births (live and still)	95% CI
<20	27	0.89	0.59 – 1.30
20 – 24	30	0.35	0.24 – 0.50
25 – 29	15	0.11	0.06 – 0.18
30 – 34	4	0.04	0.01 – 0.09
35 – 39	3	0.08	0.02 – 0.23
>40	0	0	--------
Total	**79**	**0.20**	**0.16 – 0.24**

Admittedly, the numbers are small but the fact that they have occurred in many other registries and surveillance systems makes them believable. Epidemiologic studies have not determined the cause of gastroschisis. However, some studies have found young socially disadvantaged women with a history of substance abuse to be at an increased risk. Although a

vascular insult following cocaine use has been linked, this is not proven. Gastroschisis is mostly a sporadic occurrence with a low recurrence risk however cases in siblings have been described but are rare.

In the case of prenatal diagnosis of abdominal wall defects, the risk for fetal aneuploidy in the presence of an isolated omphalocele detected by ultrasound is 40 – 60 per cent versus a negligible risk for gastroschisis. As well, omphalocele is more often associated with other fetal anomalies (50 – 70 per cent) than is gastroschisis (7 – 30 per cent).

Figure 6. Abdominal wall defects per 1000 total births, 1980 – 1998.

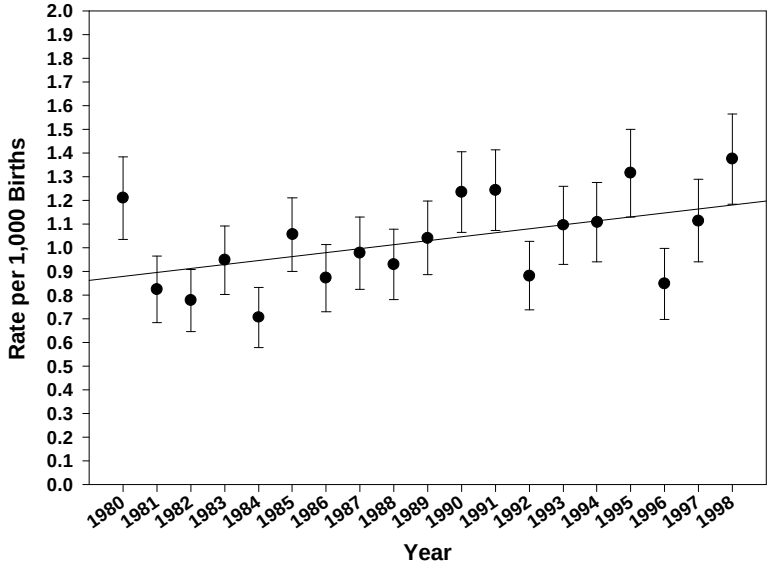


2.1.4 Down Syndrome

The vast majority of babies with Down Syndrome are the result of a non-inherited abnormality of chromosome separation during meiosis. The latter is the process of reduction division whereby the chromosome number goes from 46 to 23 in the normal process but from 46 to 24 in the abnormal process. In most instances it occurs in the mother (maternal non-disjunction) and has a definite association with increasing maternal age. The percentage of babies born to women aged 35 and over has been increasing over the last 18 years (see Appendix 8). For a variety of reasons women are deferring childbirth. Some are relying on amniocentesis followed by termination of pregnancy to avoid the consequences of advancing maternal age, however, the latter option is not acceptable to many women with the resultant increase in the provincial Down Syndrome rate.

It is also possible that there are other reasons for the increase rate of Down Syndrome such as healthier women and/or better prenatal care resulting in more live born babies with Down Syndrome which in the past might have ended either as a miscarriage or a stillbirth and thus not have been recognized as having Down Syndrome. One could also speculate that there are some environmental effects which are causing an increase in chromosomal non-disjunction but to date none has been identified.

Figure 7. Down syndrome rates per 1000 total births, 1980 – 1998.



2.1.5 Selected Other Congenital Anomalies

Figure 8. Hydrocephalus per 1000 total births, 1980 – 1998.

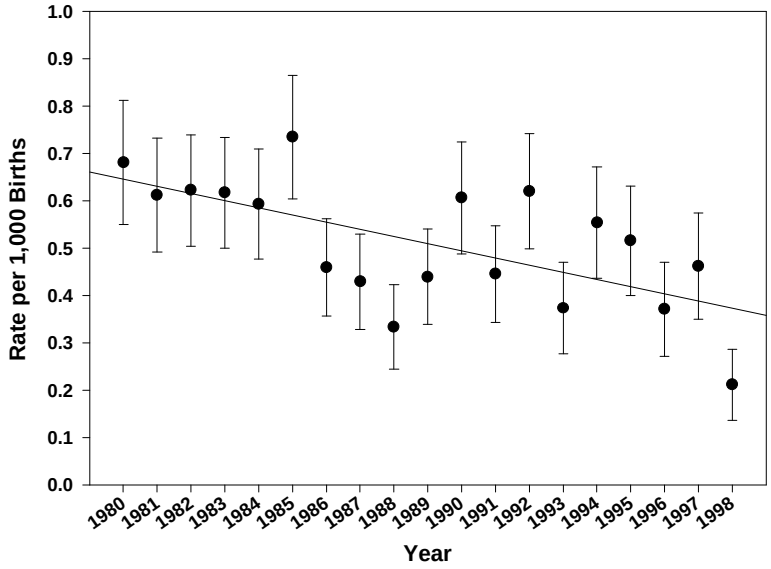


Figure 9. Limb reductions rate per 1000 total births, 1980 – 1998.

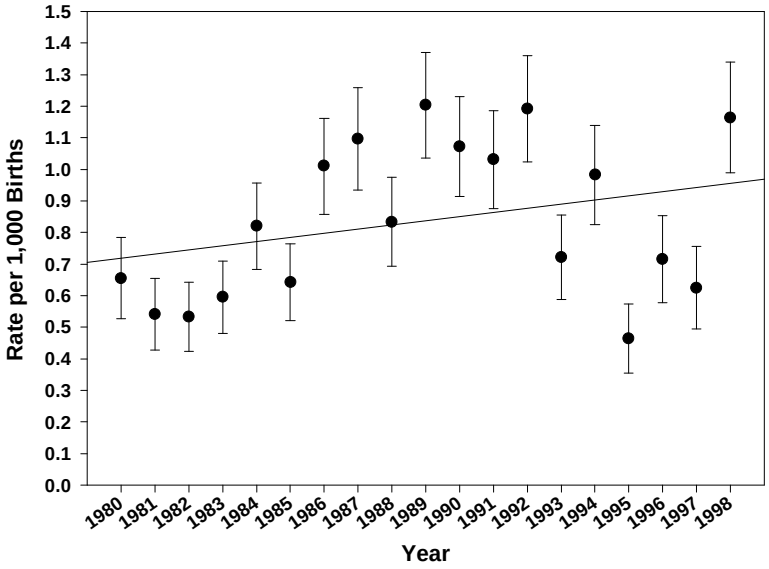


Figure 10. Renal agenesis rates per 1000 total births, 1980 –1998.

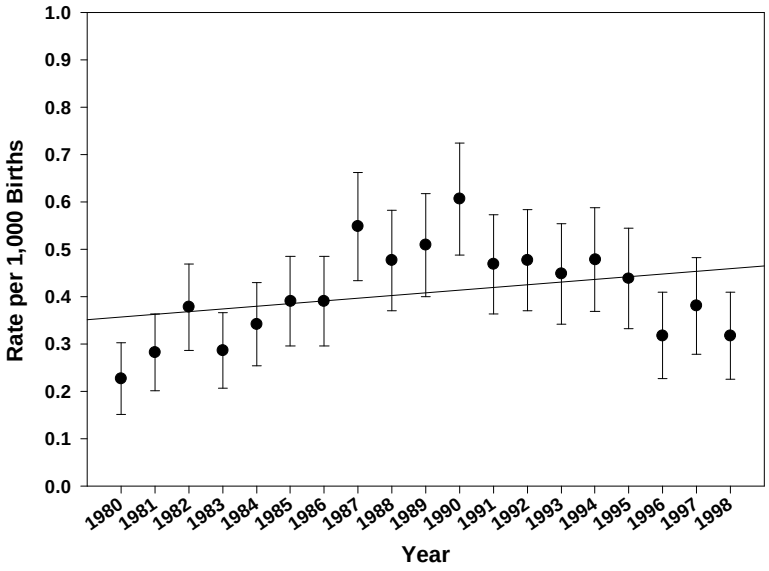


Figure 11. Hypoplastic left heart rate per 1000 total births, 1980 – 1998.

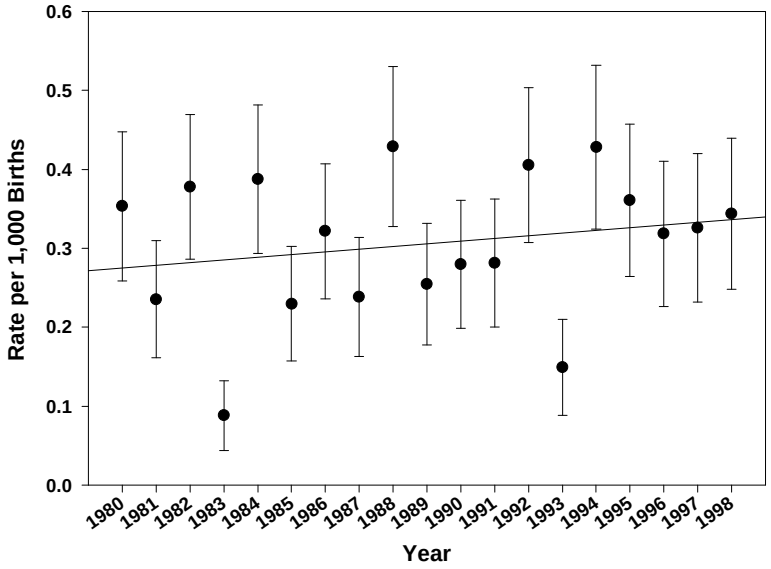


Figure 12. Esophageal atresia / stenosis rate per 1000 total births, 1980 – 1998.

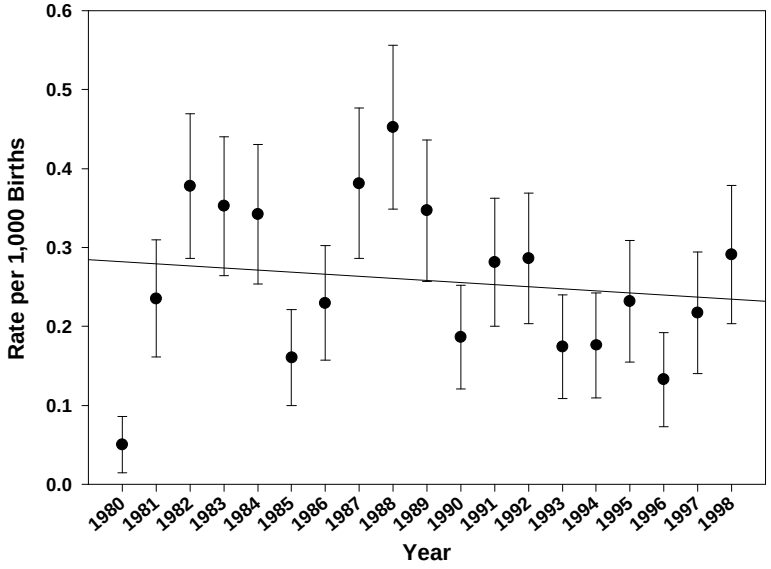


Figure 13. Anorectal atresia / stenosis rate per 1000 total births, 1980 – 1998.

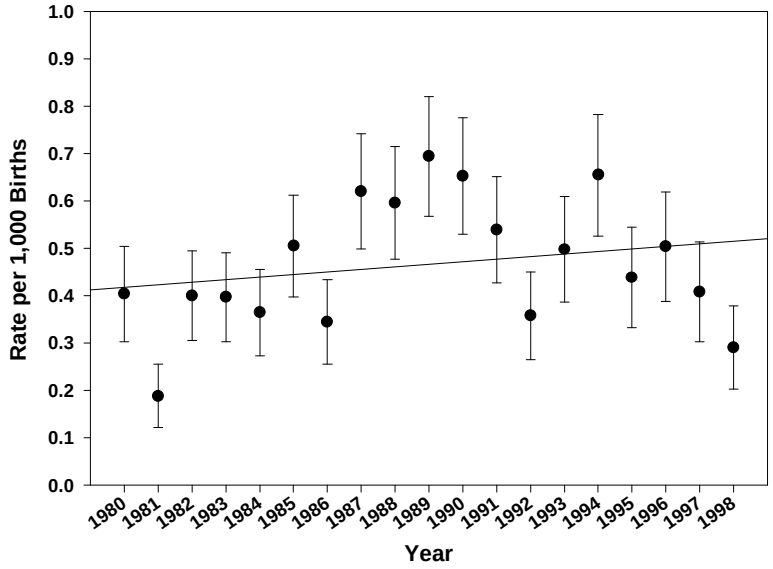
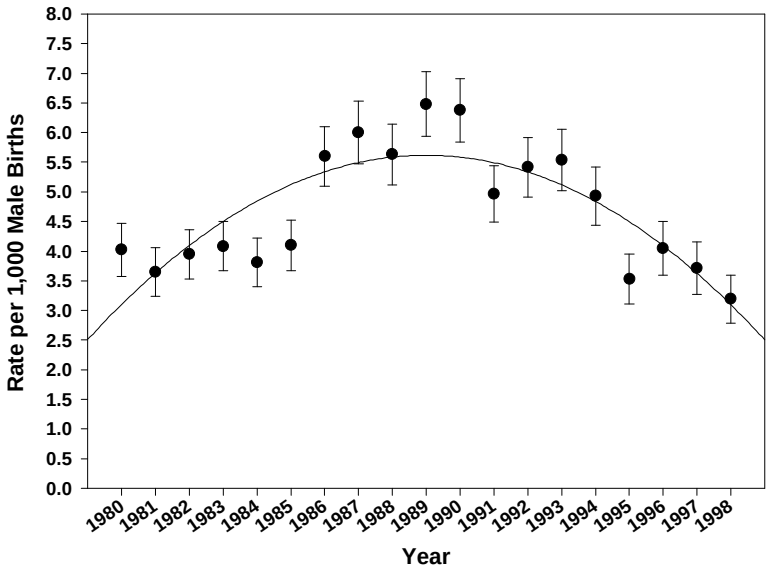


Figure 14. Hypospadias / epispadias rate per 1000 total male births, 1980 – 1998.



2.2 Baseline Rates

The baseline rates are derived from the 1985-1989 statistics whereby the rate per 1000 total births was calculated for the pooled years 1985-1989. This rate was then applied to the total births for 1996-1998 to determine an expected number of defects in the category based on the Poisson assumption.

Table 3. 1996-1998 Observed/Expected Ratios Total Births ‡

Defect	Baseline per 1000	Expected Number	Observed Number	Observed per 1000	O/E Ratio	P-Value
Anencephaly	0.32	35.94	18	0.16	0.50	0.003
Spina Bifida	0.60	67.40	43	0.38	0.63	0.003
Total NTDs	1.00	112.30	70	0.62	0.62	0.001
Hydrocephaly	0.48	53.91	39	0.38	0.79	0.042
Hypoplastic left heart	0.29	32.57	37	0.33	1.14	n.s.
Cleft Palate	0.70	78.62	89	0.79	1.13	n.s.
Cleft Lip +/- Cleft Palate	1.19	133.66	125	1.11	0.93	n.s.
Esophageal Atresia	0.31	34.82	24	0.21	0.68	0.067
Anorectal Atresia	0.55	61.77	45	0.40	0.73	0.033
Renal Agenesis/Dysgenesis	0.46	51.67	38	0.34	0.74	0.057
Hypospadias	5.56	321.50	211	1.88	0.34	0.000
Limb reductions	0.96	107.82	94	0.84	0.88	n.s.
Abdominal Wall Defects	0.43	48.30	56	0.50	1.16	n.s.
Down Syndrome	0.98	110.07	126	1.12	1.14	n.s.
Congenital Heart Defects*	8.86	995.12	987	8.79	0.99	n.s.

* includes defects found in ICD9/BPA Codes 745 and 746

‡ Total births = live births + stillbirths (terminations not included)

3.0 CONCLUSIONS

Complete prevention of birth defects is never likely to be achieved, nevertheless prospective parents can do a great deal to reduce their risks by following certain courses of action and paying attention to known risk factors. Some of these are outlined in the specific anomaly sections. General preventive measures include, knowledge of rubella antibody status, elimination of alcohol and cigarette consumption, good nutrition, and avoidance of certain medications. In particular, attention should be paid to the use of a multivitamin compound which includes folic acid which has been definitely shown to be preventive in a large number of neural tube defects and also increasing evidence that it may help to prevent orofacial clefts, limb reduction anomalies, urinary tract defects and certain forms of congenital heart disease.

Although there is great concern about potential environmental problems and pollution, we have no evidence to suggest that this is causing a change in the rates of congenital anomalies as they have remained reasonably stable over the past 18 years. Down Syndrome has increased and this is attributable to the significant increase in the per centage of live births to women aged 35 years and over. Most of the increases and decreases that we see are due to random fluctuations and/or changing ascertainment. Improvement in diagnostic techniques also can account for changes, mostly increases, e.g., congenital heart disease and renal anomalies. If any increase seems unusually suspicious we investigate these as we would any other change, e.g., geographical clustering.

In view of the change of societal attitudes with respect to termination of pregnancy it is essential to monitor the frequency of congenital anomalies in fetuses less than 20 weeks gestation. This can of course be a natural fetal death but it is more likely to be a termination of pregnancy. If these events occur after 20 weeks gestation, ascertainment will take place because the event is then registerable, i.e., a stillbirth. It is especially important to ascertain congenital anomalies in fetuses less than 20 weeks to determine the potential preventive role of multivitamins and folic acid, not only for neural tube defects but for other congenital anomalies.

Alberta is a member of the International Clearinghouse for Birth Defects Monitoring Systems (ICBDMS) and thus has access to ideas and data on congenital anomalies from nearly 50 programs representing 29 different countries. This exchange of data is an important feature and means that we do not have to wait for publications and peer reviewed journals to learn what is happening in the field of birth defects. Alberta also participates in the Group Research Studies with other members of ICBDMS.

4.0 APPENDICES

Notes:

Appendix 1	Table of Exclusion: representative of exclusions but not an exhaustive list.
Appendices 2, 3, 4, 6 & 7	Anomaly Rates: report number of anomalies, not individuals with an anomaly. These tables represent malformation counts and rates.
Appendix 5	Case Rates: report number of cases (infants) with congenital malformations regardless of the number of malformations present.

APPENDIX A1. TABLE OF EXCLUSIONS (i.e. not entered into database)

Achalasia
Adhesions – labia minora, vaginal, peritoneal
Anterior fontanelle – large, small, separated sutures
Asymmetry face, head, jaw, nose
Balanced translocation (in a normal individual with no malformations)
Birth mark, nevus or hemangioma (unless large or multiple)
Bowing of tibia or bow legs
Bronchopulmonary dysplasia
Cardiomegaly secondary to maternal diabetes
Congenital infection if no birth defects
Club foot if secondary to spina bifida
Deviated nasal septum
Diabetic cardiomyopathy
Diastasis recti
Ears – minor anomalies e.g. low set, rotated
Encephalopathy – hypoxic ischemic
Epilepsy
Esophageal reflux
Failure to thrive
Gallstones
Hairline low set
High arched palate
Heart murmurs
Hemorrhage, intracranial
Hepatomegaly/hepatosplenomegaly
Hip clicks
Hydrocele
Hydrocephalus secondary to intracranial hemorrage
Hydrops fetalis – immune (included if non-immune)
Hymen – imperforate
Hyperbilirubineamia – transient
Hypoglycemia
Intrauterine growth retardation (IUGR)
Intussusception
Isolated pulmonary hypertension
Isolated single palmar crease
Large fontanelles
Laryngeal stridor
Laryngomalacia
Leukomalacia
Mongolian spot
Nasolacrimal stenosis/blocked tear duct in a premature infant
Palsy -brachial
Patent ductus arteriosis in a premature infant or if closed within 3 months in full term infant
Patent foramen ovale in a premature infant or if closed within 3 months in full term infant
Persistent fetal circulation
Phimosis
Pilonidal dimple
Pneumothorax
Positional deformities
Ranula
Rectal prolapse
Reflux - gastroesophageal
Sacral dimple
Small anterior fontanelle
Sudden infant death syndrome (SIDS)
Sutures - overriding
Testis – retractile
Thrombocytopenia – idiopathic transient
Tongue tie
Torticollis
Tracheomalacia
Transient tyrosine elevation
Undescended testes in a premature infant
Wide suture lines
Vaginal tags

We also exclude: Infants born to Alberta non-residents and infants born out of province to Alberta residents

APPENDIX 2. SELECTED CONGENITAL ANOMALY RATES

Table A2 Selected congenital anomaly rates per 1,000 total births, 1980 – 1998, Section XIV (740.00 - 759.99).

Diagnostic Category and ICD-9/BPA Code		80-90 Subtotal	91-95 Subtotal	1996	1997	1998	91-98 Subtotal	80-98 Total
ANENCEPHALY AND SIMILAR ANOMALIES (740)	**NUMBER**	**162**	**39**	**5**	**9**	**4**	**57**	**219**
	RATE	**0.34**	**0.19**	**0.13**	**0.24**	**0.11**	**0.18**	**0.28**
	Lower CI	**0.29**	**0.14**	**0.04**	**0.11**	**0.03**	**0.14**	**0.24**
	Upper CI	**0.40**	**0.26**	**0.30**	**0.46**	**0.27**	**0.23**	**0.32**
Anencephaly (7400)	NUMBER	156	36	4	9	3	52	208
	RATE	0.33	0.18	0.11	0.24	0.08	0.16	0.26
	Lower CI	0.28	0.12	0.03	0.11	0.02	0.12	0.23
	Upper CI	0.39	0.25	0.27	0.46	0.22	0.22	0.30
Craniorachischisis (7401)	NUMBER	5	3	1	0	1	5	10
	RATE	0.01	0.01	0.03	0	0.03	0.02	0.01
	Lower CI	0.00	0.00	0.00		0.00	0.01	0.01
	Upper CI	0.02	0.04	0.13		0.13	0.04	0.02
Iniencephaly (7402)	NUMBER	1	0	0	0	0	0	1
	RATE	0.00	0	0	0	0	0	0.00
	Lower CI	0.00						0.00
	Upper CI	0.01						0.01
SPINA BIFIDA (741)	**NUMBER**	**247**	**104**	**16**	**14**	**13**	**147**	**394**
	RATE	**0.52**	**0.51**	**0.42**	**0.38**	**0.34**	**0.47**	**0.50**
	Lower CI	**0.46**	**0.42**	**0.24**	**0.21**	**0.18**	**0.39**	**0.45**
	Upper CI	**0.59**	**0.62**	**0.69**	**0.64**	**0.59**	**0.55**	**0.55**
With Hydrocephaly (7410)	NUMBER	143	60	5	4	7	76	219
	RATE	0.30	0.30	0.13	0.11	0.19	0.24	0.28
	Lower CI	0.25	0.23	0.04	0.03	0.07	0.19	0.24
	Upper CI	0.36	0.38	0.30	0.27	0.38	0.30	0.32

Number = Defects occurring in live births and stillbirths ≥ 20 weeks or ≥ 500 grams
CI = Approximate 95 per cent confidence intervals

Diagnostic Category and ICD-9/BPA Code		80-90 Subtotal	91-95 Subtotal	1996	1997	1998	91-98 Subtotal	80-98 Total
Without Hydrocephaly (7419)	NUMBER	104	44	11	10	6	73	177
	RATE	0.22	0.22	0.29	0.27	0.16	0.23	0.22
	Lower CI	0.18	0.16	0.15	0.13	0.06	0.18	0.19
	Upper CI	0.27	0.29	0.52	0.50	0.34	0.29	0.26
OTHER CONGENITAL ANOMALIES OF NERVOUS SYSTEM (742)	**NUMBER**	**727**	**337**	**58**	**58**	**54**	**507**	**1234**
	RATE	**1.54**	**1.66**	**1.54**	**1.58**	**1.43**	**1.61**	**1.56**
	Lower CI	**1.43**	**1.49**	**1.17**	**1.20**	**1.07**	**1.47**	**1.48**
	Upper CI	**1.65**	**1.84**	**1.99**	**2.04**	**1.86**	**1.75**	**1.65**
Encephalocele (7420)	NUMBER	49	28	3	2	4	37	86
	RATE	0.10	0.14	0.08	0.05	0.11	0.12	0.11
	Lower CI	0.08	0.09	0.02	0.01	0.03	0.08	0.09
	Upper CI	0.14	0.20	0.23	0.19	0.27	0.16	0.13
Microcephaly (7421)	NUMBER	145	58	6	9	8	81	226
	RATE	0.31	0.29	0.16	0.24	0.21	0.26	0.29
	Lower CI	0.26	0.22	0.06	0.11	0.09	0.20	0.25
	Upper CI	0.36	0.37	0.34	0.46	0.41	0.32	0.33
Reduction Deformities of Brain (7422)	NUMBER	121	83	18	13	11	125	246
	RATE	0.26	0.41	0.48	0.35	0.29	0.40	0.31
	Lower CI	0.21	0.33	0.28	0.19	0.15	0.33	0.27
	Upper CI	0.31	0.51	0.75	0.60	0.52	0.47	0.35
Congenital Hydrocephaly (7423)	NUMBER	261	102	14	17	8	141	402
	RATE	0.55	0.50	0.37	0.46	0.21	0.45	0.51
	Lower CI	0.49	0.41	0.20	0.27	0.09	0.38	0.46
	Upper CI	0.62	0.61	0.62	0.74	0.41	0.53	0.56
Other Specified Anomalies of Brain (7424)	NUMBER	128	59	11	15	21	106	234
	RATE	0.27	0.29	0.29	0.41	0.56	0.34	0.30
	Lower CI	0.23	0.22	0.15	0.23	0.34	0.28	0.26
	Upper CI	0.32	0.37	0.52	0.67	0.85	0.41	0.34

Number = Defects occurring in live births and stillbirths ≥ 20 weeks or ≥ 500 grams
CI = Approximate 95 per cent confidence intervals

Diagnostic Category and ICD-9/BPA Code		80-90 Subtotal	91-95 Subtotal	1996	1997	1998	91-98 Subtotal	80-98 Total
Other Specified Anomalies of Spinal Cord (7425)	NUMBER	9	2	5	2	2	11	20
	RATE	0.02	0.01	0.13	0.05	0.05	0.03	0.03
	Lower CI	0.01	0.00	0.04	0.01	0.01	0.02	0.02
	Upper CI	0.04	0.03	0.30	0.19	0.18	0.06	0.04
Other Specified Anomalies Of Nervous System (7428)	NUMBER	9	3	1	0	0	4	13
	RATE	0.02	0.01	0.03	0	0	0.01	0.02
	Lower CI	0.01	0.00	0.00			0.00	0.01
	Upper CI	0.04	0.04	0.13			0.03	0.03
Unspecified Anomalies of Brain, Spinal Cord and Nervous System (7429)	NUMBER	5	2	0	0	0	2	7
	RATE	0.01	0.01	0	0	0	0.01	0.01
	Lower CI	0.00	0.00				0.00	0.00
	Upper CI	0.02	0.03				0.02	0.02
CONGENITAL ANOMALIES OF EYE (743)	**NUMBER**	**735**	**253**	**16**	**32**	**25**	**326**	**1061**
	RATE	**1.55**	**1.24**	**0.42**	**0.87**	**0.66**	**1.03**	**1.34**
	Lower CI	**1.44**	**1.10**	**0.24**	**0.60**	**0.43**	**0.92**	**1.26**
	Upper CI	**1.67**	**1.41**	**0.69**	**1.23**	**0.98**	**1.15**	**1.43**
Anophthalmos (7430)	NUMBER	16	6	0	0	1	7	23
	RATE	0.03	0.03	0	0	0.03	0.02	0.03
	Lower CI	0.02	0.01			0.00	0.01	0.02
	Upper CI	0.05	0.06			0.13	0.05	0.04
Microphthalmos (7431)	NUMBER	47	18	1	4	3	26	73
	RATE	0.10	0.09	0.03	0.11	0.08	0.08	0.09
	Lower CI	0.07	0.05	0.00	0.03	0.02	0.05	0.07
	Upper CI	0.13	0.14	0.13	0.27	0.22	0.12	0.12
Buphthalmos (7432)	NUMBER	17	5	1	0	1	7	24
	RATE	0.04	0.02	0.03	0	0.03	0.02	0.03
	Lower CI	0.02	0.01	0.00		0.00	0.01	0.02
	Upper CI	0.06	0.06	0.13		0.13	0.05	0.05

Number = Defects occurring in live births and stillbirths ≥ 20 weeks or ≥ 500 grams
CI = Approximate 95 per cent confidence intervals

Diagnostic Category and ICD-9/BPA Code		80-90 Subtotal	91-95 Subtotal	1996	1997	1998	91-98 Subtotal	80-98 Total
Cataract and Lens Anomalies (7433)	NUMBER	62	25	5	8	5	43	105
	RATE	0.13	0.12	0.13	0.22	0.13	0.14	0.13
	Lower CI	0.10	0.08	0.04	0.09	0.04	0.10	0.11
	Upper CI	0.17	0.18	0.30	0.43	0.30	0.18	0.16
Coloboma and Other Anomalies of Anterior Segments (7434)	NUMBER	46	26	4	5	1	36	82
	RATE	0.10	0.13	0.11	0.14	0.03	0.11	0.10
	Lower CI	0.07	0.08	0.03	0.04	0.00	0.08	0.08
	Upper CI	0.13	0.19	0.27	0.31	0.13	0.16	0.13
Anomalies of Posterior Segments (7435)	NUMBER	32	10	0	2	4	16	48
	RATE	0.07	0.05	0	0.05	0.11	0.05	0.06
	Lower CI	0.05	0.02		0.01	0.03	0.03	0.04
	Upper CI	0.10	0.09		0.19	0.27	0.08	0.08
Anomalies of Eyelids, Lacrimal System and Orbit (7436)	NUMBER	484	150	5	12	6	173	657
	RATE	1.02	0.74	0.13	0.33	0.16	0.55	0.83
	Lower CI	0.93	0.62	0.04	0.17	0.06	0.47	0.77
	Upper CI	1.12	0.87	0.30	0.57	0.34	0.64	0.90
Other Specified Anomalies of Eye (7438)	NUMBER	26	13	0	1	4	18	44
	RATE	0.05	0.06	0	0.03	0.11	0.06	0.06
	Lower CI	0.04	0.03		0.00	0.03	0.03	0.04
	Upper CI	0.08	0.11		0.14	0.27	0.09	0.07
Unspecified Anomalies of Eye (7439)	NUMBER	5	0	0	0	0	0	5
	RATE	0.01	0	0	0	0	0	0.01
	Lower CI	0.00						0.00
	Upper CI	0.02						0.01
CONGENITAL ANOMALIES OF EAR, FACE AND NECK (744)	**NUMBER**	**1337**	**577**	**82**	**84**	**100**	**843**	**2180**
	RATE	**2.82**	**2.84**	**2.17**	**2.28**	**2.64**	**2.67**	**2.76**
	Lower CI	**2.67**	**2.61**	**1.73**	**1.82**	**2.15**	**2.49**	**2.65**
	Upper CI	**2.98**	**3.08**	**2.70**	**2.83**	**3.22**	**2.86**	**2.88**

Number = Defects occurring in live births and stillbirths ≥ 20 weeks or ≥ 500 grams
CI = Approximate 95 per cent confidence intervals

Diagnostic Category and ICD-9/BPA Code		80-90 Subtotal	91-95 Subtotal	1996	1997	1998	91-98 Subtotal	80-98 Total
Anomalies of Ear Causing Hearing Impairment (7440)	NUMBER	46	24	4	3	7	38	84
	RATE	0.10	0.12	0.11	0.08	0.19	0.12	0.11
	Lower CI	0.07	0.08	0.03	0.02	0.07	0.09	0.08
	Upper CI	0.13	0.18	0.27	0.23	0.38	0.17	0.13
Accessory Auricle (7441)	NUMBER	900	385	59	57	68	569	1469
	RATE	1.90	1.89	1.56	1.55	1.80	1.80	1.86
	Lower CI	1.78	1.71	1.19	1.17	1.40	1.66	1.77
	Upper CI	2.03	2.09	2.02	2.01	2.28	1.96	1.96
Other Specified Anomalies of Ear (7442)	NUMBER	149	79	8	7	12	106	255
	RATE	0.31	0.39	0.21	0.19	0.32	0.34	0.32
	Lower CI	0.27	0.31	0.09	0.08	0.16	0.28	0.28
	Upper CI	0.37	0.48	0.41	0.39	0.55	0.41	0.37
Unspecified Anomalies of Ear (7443)	NUMBER	76	21	0	3	2	26	102
	RATE	0.16	0.10	0	0.08	0.05	0.08	0.13
	Lower CI	0.13	0.06		0.02	0.01	0.05	0.11
	Upper CI	0.20	0.16		0.23	0.18	0.12	0.16
Branchial Cleft, Cyst or Fistula Preauricular Sinus (7444)	NUMBER	132	57	9	12	7	85	217
	RATE	0.28	0.28	0.24	0.33	0.19	0.27	0.27
	Lower CI	0.23	0.21	0.11	0.17	0.07	0.22	0.24
	Upper CI	0.33	0.36	0.45	0.57	0.38	0.33	0.31
Webbing of Neck (7445)	NUMBER	12	2	2	1	1	6	18
	RATE	0.03	0.01	0.05	0.03	0.03	0.02	0.02
	Lower CI	0.01	0.00	0.01	0.00	0.00	0.01	0.01
	Upper CI	0.04	0.03	0.18	0.14	0.13	0.04	0.04
Other Specified Anomalies of Face and Neck (7448)	NUMBER	15	9	0	1	3	13	28
	RATE	0.03	0.04	0	0.03	0.08	0.04	0.04
	Lower CI	0.02	0.02		0.00	0.02	0.02	0.02
	Upper CI	0.05	0.08		0.14	0.22	0.07	0.05

Number = Defects occurring in live births and stillbirths ≥ 20 weeks or ≥ 500 grams
CI = Approximate 95 per cent confidence intervals

Diagnostic Category and ICD-9/BPA Code		80-90 Subtotal	91-95 Subtotal	1996	1997	1998	91-98 Subtotal	80-98 Total
Unspecified Anomalies of Face and Neck (7449)	NUMBER	7	0	0	0	0	0	7
	RATE	0.01	0	0	0	0	0	0.01
	Lower CI	0.01						0.00
	Upper CI	0.03						0.02
BULBUS CORDIS ANOMALIES AND ANOMALIES OF CARDIAC SEPTAL CLOSURE (745)	**NUMBER**	**2560**	**1222**	**232**	**212**	**197**	**1863**	**4423**
	RATE	**5.41**	**6.01**	**6.15**	**5.76**	**5.21**	**5.90**	**5.60**
	Lower CI	**5.20**	**5.68**	**5.39**	**5.01**	**4.51**	**5.64**	**5.44**
	Upper CI	**5.62**	**6.36**	**7.00**	**6.59**	**5.99**	**6.18**	**5.77**
Common Truncus (7450)	NUMBER	44	13	3	3	5	24	68
	RATE	0.09	0.06	0.08	0.08	0.13	0.08	0.09
	Lower CI	0.07	0.03	0.02	0.02	0.04	0.05	0.07
	Upper CI	0.12	0.11	0.23	0.23	0.30	0.11	0.11
Transposition of Great Vessels (7451)	NUMBER	142	55	16	15	9	95	237
	RATE	0.30	0.27	0.42	0.41	0.24	0.30	0.30
	Lower CI	0.25	0.20	0.24	0.23	0.11	0.24	0.26
	Upper CI	0.35	0.35	0.69	0.67	0.45	0.37	0.34
Tetralogy of Fallot (7452)	NUMBER	104	61	11	7	7	86	190
	RATE	0.22	0.30	0.29	0.19	0.19	0.27	0.24
	Lower CI	0.18	0.23	0.15	0.08	0.07	0.22	0.21
	Upper CI	0.27	0.39	0.52	0.39	0.38	0.34	0.28
Common Ventricle (7453)	NUMBER	51	36	12	4	8	60	111
	RATE	0.11	0.18	0.32	0.11	0.21	0.19	0.14
	Lower CI	0.08	0.12	0.16	0.03	0.09	0.15	0.12
	Upper CI	0.14	0.25	0.55	0.27	0.41	0.24	0.17
Ventricular Septal Defect (7454)	NUMBER	1373	576	103	80	95	854	2227
	RATE	2.90	2.83	2.73	2.17	2.51	2.71	2.82
	Lower CI	2.75	2.61	2.23	1.72	2.03	2.53	2.71
	Upper CI	3.06	3.08	3.31	2.71	3.07	2.89	2.94

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Diagnostic Category and ICD-9/BPA Code		80-90 Subtotal	91-95 Subtotal	1996	1997	1998	91-98 Subtotal	80-98 Total
Ostium Secundum Type Atrial Septal Defect (7455)	NUMBER	680	420	75	91	58	644	1324
	RATE	1.44	2.07	1.99	2.47	1.53	2.04	1.68
	Lower CI	1.33	1.87	1.57	1.99	1.17	1.89	1.59
	Upper CI	1.55	2.27	2.49	3.04	1.98	2.20	1.77
Endocardial Cushion Defects (7456)	NUMBER	155	61	12	12	15	100	255
	RATE	0.33	0.30	0.32	0.33	0.40	0.32	0.32
	Lower CI	0.28	0.23	0.16	0.17	0.22	0.26	0.28
	Upper CI	0.38	0.39	0.55	0.57	0.65	0.39	0.37
Cor Biloculare (7457)	NUMBER	3	0	0	0	0	0	3
	RATE	0.01	0	0	0	0	0	0.00
	Lower CI	0.00						0.00
	Upper CI	0.02						0.01
Other (7458)	NUMBER	5	0	0	0	0	0	5
	RATE	0.01	0	0	0	0	0	0.01
	Lower CI	0.00						0.00
	Upper CI	0.02						0.01
Unspecified Defect of Septal Closure (7459)	NUMBER	3	0	0	0	0	0	3
	RATE	0.01	0	0	0	0	0	0.00
	Lower CI	0.00						0.00
	Upper CI	0.02						0.01
OTHER CONGENITAL ANOMALIES OF THE HEART (746)	**NUMBER**	**1125**	**503**	**118**	**115**	**113**	**849**	**1974**
	RATE	**2.38**	**2.47**	**3.13**	**3.13**	**2.99**	**2.69**	**2.50**
	Lower CI	**2.24**	**2.26**	**2.59**	**2.58**	**2.46**	**2.51**	**2.39**
	Upper CI	**2.52**	**2.70**	**3.75**	**3.75**	**3.59**	**2.88**	**2.61**
Anomalies of Pulmonary Valve (7460)	NUMBER	381	130	30	27	28	215	596
	RATE	0.80	0.64	0.80	0.73	0.74	0.68	0.76
	Lower CI	0.73	0.53	0.54	0.48	0.49	0.59	0.70
	Upper CI	0.89	0.76	1.14	1.07	1.07	0.78	0.82

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Diagnostic Category and ICD-9/BPA Code		80-90 Subtotal	91-95 Subtotal	1996	1997	1998	91-98 Subtotal	80-98 Total
Tricuspid Atresia and Stenosis (7461)	NUMBER	87	81	13	21	15	130	217
	RATE	0.18	0.40	0.34	0.57	0.40	0.41	0.27
	Lower CI	0.15	0.32	0.18	0.35	0.22	0.34	0.24
	Upper CI	0.23	0.50	0.59	0.87	0.65	0.49	0.31
Ebstein's Anomaly (7462)	NUMBER	22	9	0	2	0	11	33
	RATE	0.05	0.04	0	0.05	0	0.03	0.04
	Lower CI	0.03	0.02		0.01		0.02	0.03
	Upper CI	0.07	0.08		0.19		0.06	0.06
Stenosis of Aortic Valve (7463)	NUMBER	56	31	12	10	5	58	114
	RATE	0.12	0.15	0.32	0.27	0.13	0.18	0.14
	Lower CI	0.09	0.10	0.16	0.13	0.04	0.14	0.12
	Upper CI	0.15	0.22	0.55	0.50	0.30	0.24	0.17
Insufficiency of Aortic Valve (7464)	NUMBER	55	33	12	9	19	73	128
	RATE	0.12	0.16	0.32	0.21	0.50	0.23	0.16
	Lower CI	0.09	0.11	0.16	0.11	0.30	0.18	0.14
	Upper CI	0.15	0.23	0.55	0.46	0.78	0.29	0.19
Mitral Stenosis (7465)	NUMBER	31	4	2	6	4	16	47
	RATE	0.07	0.02	0.05	0.16	0.11	0.05	0.06
	Lower CI	0.04	0.01	0.01	0.06	0.03	0.03	0.04
	Upper CI	0.09	0.05	0.18	0.35	0.27	0.08	0.08
Mitral Insufficiency (7466)	NUMBER	21	30	7	10	10	57	78
	RATE	0.04	0.15	0.19	0.27	0.26	0.18	0.10
	Lower CI	0.03	0.10	0.07	0.13	0.13	0.14	0.08
	Upper CI	0.07	0.21	0.38	0.50	0.48	0.23	0.12
Hypoplastic Left Heart Syndrome (7467)	NUMBER	140	65	12	12	13	102	242
	RATE	0.30	0.32	0.32	0.33	0.34	0.32	0.31
	Lower CI	0.25	0.25	0.16	0.17	0.18	0.26	0.27
	Upper CI	0.35	0.41	0.55	0.57	0.59	0.39	0.35

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Diagnostic Category and ICD-9/BPA Code		80-90 Subtotal	91-95 Subtotal	1996	1997	1998	91-98 Subtotal	80-98 Total
Other Specified Anomalies of Heart (7468)	NUMBER	262	116	30	18	18	182	444
	RATE	0.55	0.57	0.80	0.49	0.48	0.58	0.56
	Lower CI	0.49	0.47	0.54	0.29	0.28	0.50	0.51
	Upper CI	0.62	0.68	1.14	0.77	0.75	0.67	0.62
Unspecified Anomalies of Heart (7469)	NUMBER	70	4	0	0	1	5	75
	RATE	0.15	0.02	0	0	0.03	0.02	0.10
	Lower CI	0.12	0.01			0.00	0.01	0.07
	Upper CI	0.19	0.05			0.13	0.04	0.12
OTHER CONGENITAL ANOMALIES OF CIRCULATORY SYSTEM (747)	**NUMBER**	**2411**	**827**	**113**	**122**	**127**	**1189**	**3600**
	RATE	**5.09**	**4.07**	**3.00**	**3.32**	**3.36**	**3.77**	**4.56**
	Lower CI	**4.89**	**3.80**	**2.47**	**2.75**	**2.80**	**3.56**	**4.41**
	Upper CI	**5.30**	**4.36**	**3.60**	**3.96**	**4.00**	**3.99**	**4.71**
Patent Ductus Arteriosus (7470)	NUMBER	1158	188	21	28	17	254	1412
	RATE	2.45	0.92	0.56	0.76	0.45	0.80	1.79
	Lower CI	2.31	0.80	0.35	0.51	0.26	0.71	1.70
	Upper CI	2.59	1.07	0.85	1.10	0.72	0.91	1.89
Coarctation of the Aorta (7471)	NUMBER	187	108	23	18	12	161	348
	RATE	0.39	0.53	0.61	0.49	0.32	0.51	0.44
	Lower CI	0.34	0.44	0.39	0.29	0.16	0.43	0.40
	Upper CI	0.46	0.64	0.91	0.77	0.55	0.60	0.49
Anomalies of Aorta (7472)	NUMBER	66	21	3	7	8	39	105
	RATE	0.14	0.10	0.08	0.19	0.21	0.12	0.13
	Lower CI	0.11	0.06	0.02	0.08	0.09	0.09	0.11
	Upper CI	0.18	0.16	0.23	0.39	0.41	0.17	0.16
Anomalies of Pulmonary Artery (7473)	NUMBER	136	69	8	13	9	99	235
	RATE	0.29	0.34	0.21	0.35	0.24	0.31	0.30
	Lower CI	0.24	0.26	0.09	0.19	0.11	0.26	0.26
	Upper CI	0.34	0.43	0.41	0.60	0.45	0.38	0.34

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Diagnostic Category and ICD-9/BPA Code		80-90 Subtotal	91-95 Subtotal	1996	1997	1998	91-98 Subtotal	80-98 Total
Anomalies of Great Veins (7474)	NUMBER	87	39	8	3	15	65	152
	RATE	0.18	0.19	0.21	0.08	0.40	0.21	0.19
	Lower CI	0.15	0.14	0.09	0.02	0.22	0.16	0.16
	Upper CI	0.23	0.26	0.41	0.23	0.65	0.26	0.23
Absence or Hypoplasia of Umbilical Artery (7475)	NUMBER	489	309	40	46	56	451	940
	RATE	1.03	1.52	1.06	1.25	1.48	1.43	1.19
	Lower CI	0.94	1.36	0.76	0.92	1.12	1.30	1.12
	Upper CI	1.13	1.70	1.44	1.67	1.92	1.57	1.27
Other Anomalies of Peripheral Vascular System (7476)	NUMBER	24	23	3	3	6	35	59
	RATE	0.05	0.11	0.08	0.08	0.16	0.11	0.07
	Lower CI	0.03	0.07	0.02	0.02	0.06	0.08	0.06
	Upper CI	0.08	0.17	0.23	0.23	0.34	0.15	0.10
Other Specified Anomalies of Circulatory System (7478)	NUMBER	261	72	7	4	4	87	348
	RATE	0.55	0.35	0.19	0.11	0.11	0.28	0.44
	Lower CI	0.49	0.28	0.07	0.03	0.03	0.22	0.40
	Upper CI	0.62	0.45	0.38	0.27	0.27	0.34	0.49
Unspecified Anomalies of Circulatory System (7479)	NUMBER	3	0	0	0	0	0	3
	RATE	0.01	0	0	0	0	0	0.00
	Lower CI	0.00						0.00
	Upper CI	0.02						0.01
CONGENITAL ANOMALIES OF RESPIRATORY SYSTEM (748)	**NUMBER**	**367**	**110**	**22**	**13**	**22**	**167**	**534**
	RATE	**0.78**	**0.54**	**0.58**	**0.35**	**0.58**	**0.53**	**0.68**
	Lower CI	**0.70**	**0.44**	**0.37**	**0.19**	**0.37**	**0.45**	**0.62**
	Upper CI	**0.86**	**0.65**	**0.88**	**0.60**	**0.88**	**0.62**	**0.74**
Choanal Atresia (7480)	NUMBER	65	35	12	4	1	52	117
	RATE	0.14	0.17	0.32	0.11	0.03	0.16	0.15
	Lower CI	0.11	0.12	0.16	0.03	0.00	0.12	0.12
	Upper CI	0.17	0.24	0.55	0.27	0.13	0.22	0.18

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Diagnostic Category and ICD-9/BPA Code		80-90 Subtotal	91-95 Subtotal	1996	1997	1998	91-98 Subtotal	80-98 Total
Other Anomalies of Nose (7481)	NUMBER	25	9	2	2	1	14	39
	RATE	0.05	0.04	0.05	0.05	0.03	0.04	0.05
	Lower CI	0.03	0.02	0.01	0.01	0.00	0.02	0.04
	Upper CI	0.08	0.08	0.18	0.19	0.13	0.07	0.07
Web of Larynx (7482)	NUMBER	8	1	0	0	1	2	10
	RATE	0.02	0.00	0	0	0.03	0.01	0.01
	Lower CI	0.01	0.00			0.00	0.00	0.01
	Upper CI	0.03	0.02			0.13	0.02	0.02
Other Anomalies of Larynx, Trachea and Bronchus (7483)	NUMBER	45	18	2	3	7	30	75
	RATE	0.10	0.09	0.05	0.08	0.19	0.10	0.10
	Lower CI	0.07	0.05	0.01	0.02	0.07	0.06	0.07
	Upper CI	0.13	0.14	0.18	0.23	0.38	0.14	0.12
Cystic Lung (7484)	NUMBER	10	11	0	1	5	17	27
	RATE	0.02	0.05	0	0.03	0.13	0.05	0.03
	Lower CI	0.01	0.03		0.00	0.04	0.03	0.02
	Upper CI	0.04	0.10		0.14	0.30	0.09	0.05
Agenesis, Hypoplasia and Dysplasia of Lung (7485)	NUMBER	196	28	4	1	7	40	236
	RATE	0.41	0.14	0.11	0.03	0.19	0.13	0.30
	Lower CI	0.36	0.09	0.03	0.00	0.07	0.09	0.26
	Upper CI	0.48	0.20	0.27	0.14	0.38	0.17	0.34
Other Anomalies of Lung (7486)	NUMBER	15	4	2	2	0	8	23
	RATE	0.03	0.02	0.05	0.05	0	0.03	0.03
	Lower CI	0.02	0.01	0.01	0.01		0.01	0.02
	Upper CI	0.05	0.05	0.18	0.19		0.05	0.04
Other Specified Anomalies of Respiratory System (7488)	NUMBER	3	4	0	0	0	4	7
	RATE	0.01	0.02	0	0	0	0.01	0.01
	Lower CI	0.00	0.01				0.00	0.00
	Upper CI	0.02	0.05				0.03	0.02

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Diagnostic Category and ICD-9/BPA Code		80-90 Subtotal	91-95 Subtotal	1996	1997	1998	91-98 Subtotal	80-98 Total
CLEFT PALATE AND CLEFT LIP (749)	**NUMBER**	**862**	**387**	**68**	**71**	**75**	**601**	**1436**
	RATE	**1.82**	**1.90**	**1.80**	**1.93**	**1.98**	**1.90**	**1.85**
	Lower CI	**1.70**	**1.72**	**1.40**	**1.51**	**1.56**	**1.76**	**1.76**
	Upper CI	**1.95**	**2.10**	**2.28**	**2.43**	**2.49**	**2.06**	**1.95**
Cleft Palate (7490)	NUMBER	333	154	23	38	28	243	576
	RATE	0.70	0.76	0.61	1.03	0.74	0.77	0.73
	Lower CI	0.63	0.64	0.39	0.73	0.49	0.68	0.67
	Upper CI	0.78	0.89	0.91	1.42	1.07	0.87	0.79
Cleft Lip (7491)	NUMBER	163	90	19	13	19	141	304
	RATE	0.34	0.44	0.50	0.35	0.50	0.45	0.39
	Lower CI	0.29	0.36	0.30	0.19	0.30	0.38	0.34
	Upper CI	0.40	0.54	0.79	0.60	0.78	0.53	0.43
Cleft Palate with Cleft Lip (7492)	NUMBER	366	143	26	20	28	217	583
	RATE	0.77	0.70	0.69	0.54	0.74	0.69	0.74
	Lower CI	0.80	0.59	0.45	0.33	0.49	0.60	0.68
	Upper CI	0.76	0.83	1.01	0.84	1.07	0.79	0.80
OTHER CONGENITAL ANOMALIES OF UPPER ALIMENTARY TRACT (750)	**NUMBER**	**640**	**217**	**29**	**37**	**43**	**326**	**966**
	RATE	**1.35**	**1.07**	**0.77**	**1.01**	**1.14**	**1.03**	**1.22**
	Lower CI	**1.25**	**0.93**	**0.52**	**0.71**	**0.82**	**0.92**	**1.15**
	Upper CI	**1.46**	**1.22**	**1.10**	**1.39**	**1.53**	**1.15**	**1.30**
Other Anomalies of Tongue (7501)	NUMBER	31	16	1	0	3	20	51
	RATE	0.07	0.08	0.03	0	0.08	0.06	0.06
	Lower CI	0.04	0.05	0.00		0.02	0.04	0.05
	Upper CI	0.09	0.13	0.13		0.22	0.10	0.09
Other Specified Anomalies of Mouth And Pharynx (7502)	NUMBER	21	18	1	2	3	24	45
	RATE	0.04	0.09	0.03	0.05	0.08	0.08	0.06
	Lower CI	0.03	0.05	0.00	0.01	0.02	0.05	0.04
	Upper CI	0.07	0.14	0.13	0.19	0.22	0.11	0.08

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Diagnostic Category and ICD-9/BPA Code		80-90 Subtotal	91-95 Subtotal	1996	1997	1998	91-98 Subtotal	80-98 Total
Tracheo-Esophageal Fistula, Esophageal Atresia and Stenosis (7503)	NUMBER	135	46	5	8	11	70	205
	RATE	0.29	0.23	0.13	0.22	0.29	0.22	0.26
	Lower CI	0.24	0.17	0.04	0.09	0.15	0.17	0.23
	Upper CI	0.34	0.30	0.30	0.43	0.52	0.28	0.30
Other Specified Anomalies of Esophagus (7504)	NUMBER	2	1	0	0	0	1	3
	RATE	0.00	0.00	0	0	0	0.00	0.00
	Lower CI	0.00	0.00				0.00	0.00
	Upper CI	0.01	0.02				0.02	0.01
Hypertrophic Pyloric Stenosis (7505)	NUMBER	441	134	22	26	25	207	648
	RATE	0.93	0.66	0.58	0.71	0.66	0.66	0.82
	Lower CI	0.85	0.55	0.37	0.46	0.43	0.57	0.76
	Upper CI	1.02	0.78	0.88	1.04	0.98	0.75	0.89
Hiatus Hernia (7506)	NUMBER	5	2	0	0	1	3	8
	RATE	0.01	0.01	0	0	0.03	0.01	0.01
	Lower CI	0.00	0.00			0.00	0.00	0.00
	Upper CI	0.02	0.03			0.13	0.03	0.02
Other Specified Anomalies of Stomach (7507)	NUMBER	4	0	0	0	0	0	4
	RATE	0.01	0	0	0	0	0	0.01
	Lower CI	0.00						0.00
	Upper CI	0.02						0.01
Unspecified Anomalies of Upper Alimentary Tract (7509)	NUMBER	1	0	0	0	0	0	1
	RATE	0.00	0	0	0	0	0	0.00
	Lower CI	0.00						0.00
	Upper CI	0.01						0.01
OTHER CONGENITAL ANOMALIES OF THE DIGESTIVE SYSTEM (751)	**NUMBER**	**703**	**315**	**64**	**60**	**72**	**511**	**1214**
	RATE	**1.48**	**1.55**	**1.70**	**1.63**	**1.90**	**1.62**	**1.54**
	Lower CI	**1.38**	**1.38**	**1.31**	**1.25**	**1.49**	**1.48**	**1.45**
	Upper CI	**1.60**	**1.73**	**2.17**	**2.10**	**2.40**	**1.77**	**1.63**

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Diagnostic Category and ICD-9/BPA Code		80-90 Subtotal	91-95 Subtotal	1996	1997	1998	91-98 Subtotal	80-98 Total
Meckel's Diverticulum (7510)	NUMBER	52	13	4	2	3	22	74
	RATE	0.11	0.06	0.11	0.05	0.08	0.07	0.09
	Lower CI	0.08	0.03	0.03	0.01	0.02	0.04	0.07
	Upper CI	0.14	0.11	0.27	0.19	0.22	0.11	0.12
Atresia and Stenosis of Small Intestine (7511)	NUMBER	105	50	9	10	11	80	185
	RATE	0.22	0.25	0.24	0.27	0.29	0.25	0.23
	Lower CI	0.18	0.18	0.11	0.13	0.15	0.20	0.20
	Upper CI	0.27	0.32	0.45	0.50	0.52	0.32	0.27
Atresia and Stenosis of Large Intestine, Rectum, and Anal Canal (7512)	NUMBER	219	100	19	15	11	145	366
	RATE	0.46	0.49	0.50	0.41	0.29	0.46	0.46
	Lower CI	0.40	0.40	0.30	0.23	0.15	0.39	0.42
	Upper CI	0.53	0.60	0.79	0.67	0.52	0.54	0.51
Hirschsprung's Disease and Other Functional Disorders of Colon (7513)	NUMBER	68	33	4	4	4	45	113
	RATE	0.14	0.16	0.11	0.11	0.11	0.14	0.14
	Lower CI	0.11	0.11	0.03	0.03	0.03	0.10	0.12
	Upper CI	0.18	0.23	0.27	0.27	0.27	0.19	0.17
Anomalies of Intestinal Fixation (7514)	NUMBER	122	44	13	13	27	97	219
	RATE	0.26	0.22	0.34	0.35	0.71	0.31	0.28
	Lower CI	0.21	0.16	0.18	0.19	0.47	0.25	0.24
	Upper CI	0.31	0.29	0.59	0.60	1.04	0.37	0.32
Other Anomalies of Intestine (7515)	NUMBER	75	45	9	10	7	71	146
	RATE	0.16	0.22	0.24	0.27	0.19	0.22	0.19
	Lower CI	0.12	0.16	0.11	0.13	0.07	0.18	0.16
	Upper CI	0.20	0.30	0.45	0.50	0.38	0.28	0.22
Anomalies of Gall Bladder, Bile Ducts and Liver (7516)	NUMBER	47	24	4	5	6	39	86
	RATE	0.10	0.12	0.11	0.14	0.16	0.12	0.11
	Lower CI	0.07	0.08	0.03	0.04	0.06	0.09	0.09
	Upper CI	0.13	0.18	0.27	0.31	0.34	0.17	0.13

Number = Defects occurring in live births and stillbirths ≥ 20 weeks or ≥ 500 grams
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Diagnostic Category and ICD-9/BPA Code		80-90 Subtotal	91-95 Subtotal	1996	1997	1998	91-98 Subtotal	80-98 Total
Anomalies of Pancreas (7517)	NUMBER	11	7	1	1	3	12	23
	RATE	0.02	0.03	0.03	0.03	0.08	0.04	0.03
	Lower CI	0.01	0.01	0.00	0.00	0.02	0.02	0.02
	Upper CI	0.04	0.07	0.13	0.14	0.22	0.07	0.04
Other Specified Anomalies of Digestive System (7518)	NUMBER	2	0	1	0	0	1	3
	RATE	0.00	0	0.03	0	0	0.00	0.00
	Lower CI	0.00		0.00			0.00	0.00
	Upper CI	0.01		0.13			0.02	0.01
Unspecified Anomalies of Digestive System (7519)	NUMBER	2	0	0	0	0	0	2
	RATE	0.00	0	0	0	0	0	0.00
	Lower CI	0.00						0.00
	Upper CI	0.01						0.01
CONGENITAL ANOMALIES OF GENITAL ORGANS (752)	**NUMBER**	**2759**	**1163**	**191**	**184**	**191**	**1729**	**4488**
	RATE	**5.83**	**5.72**	**5.06**	**5.00**	**5.05**	**5.48**	**5.69**
	Lower CI	**5.61**	**5.40**	**4.37**	**4.30**	**4.36**	**5.22**	**5.52**
	Upper CI	**6.05**	**6.06**	**5.84**	**5.78**	**5.82**	**5.74**	**5.86**
Anomalies of Ovaries (7520) *	NUMBER	14	2	0	0	3	5	19
	RATE	0.06	0.02	0	0	0.16	0.03	0.05
	Lower CI	0.03	0.00			0.03	0.01	0.03
	Upper CI	0.10	0.07			0.46	0.07	0.08
Anomalies of Fallopian Tubes and Broad Ligaments (7521) *	NUMBER	7	0	0	1	0	1	8
	RATE	0.03	0	0	0.06	0	0.01	0.02
	Lower CI	0.01			0.00		0.00	0.01
	Upper CI	0.06			0.28		0.03	0.04
Doubling of Uterus (7522) *	NUMBER	3	0	0	0	0	0	3
	RATE	0.01	0	0	0	0	0	0.01
	Lower CI	0.00						0.00
	Upper CI	0.04						0.02

* Rates based on female births only

Number = Defects occurring in live births and stillbirths ≥ 20 weeks or ≥ 500 grams

CI = Approximate 95 per cent confidence intervals

Diagnostic Category and ICD-9/BPA Code		80-90 Subtotal	91-95 Subtotal	1996	1997	1998	91-98 Subtotal	80-98 Total
Other Anomalies of Uterus (7523) *	NUMBER	21	12	0	2	3	17	38
	RATE	0.09	0.12	0	0.11	0.16	0.11	0.10
	Lower CI	0.06	0.06		0.01	0.03	0.06	0.07
	Upper CI	0.14	0.21		0.38	0.46	0.18	0.14
Anomalies of Cervix, Vagina and External Female Genitalia (7524)*	NUMBER	36	12	2	2	0	16	52
	RATE	0.16	0.12	0.11	0.11	0	0.10	0.14
	Lower CI	0.11	0.06	0.01	0.01		0.06	0.10
	Upper CI	0.22	0.21	0.38	0.38		0.17	0.18
Undescended Testicle (7525) **	NUMBER	1274	505	88	90	105	788	2062
	RATE	5.28	4.81	4.51	4.77	5.40	4.84	5.10
	Lower CI	4.99	4.40	3.62	3.84	4.42	4.51	4.88
	Upper CI	5.58	5.25	5.55	5.87	6.54	5.19	5.33
Hypospadias and Epispadias (7526)**	NUMBER	1178	508	79	70	62	719	1897
	RATE	4.88	4.84	4.05	3.71	3.19	4.42	4.69
	Lower CI	4.61	4.43	3.21	2.89	2.45	4.10	4.49
	Upper CI	5.17	5.28	5.04	4.69	4.09	4.75	4.91
Indeterminate Sex and Pseudohermaphroditism (7527)	NUMBER	24	23	1	2	3	29	53
	RATE	0.05	0.11	0.03	0.05	0.08	0.09	0.07
	Lower CI	0.03	0.07	0.00	0.01	0.02	0.06	0.05
	Upper CI	0.08	0.17	0.13	0.19	0.22	0.13	0.09
Other Specified Anomalies of Male Genital Organs (7528)**	NUMBER	199	101	21	17	15	154	353
	RATE	0.82	0.96	1.08	0.90	0.77	0.95	0.87
	Lower CI	0.71	0.78	0.67	0.53	0.43	0.80	0.78
	Upper CI	0.95	1.17	1.64	1.44	1.27	1.11	0.97
Unspecified Anomalies of Genital Organs (7529)	NUMBER	3	0	0	0	0	0	3
	RATE	0.01	0	0	0	0	0	0.00
	Lower CI	0.00						0.00
	Upper CI	0.02						0.01

* Rates based on female births only

** Rates based on male births only

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CI = Approximate 95 per cent confidence intervals

Diagnostic Category and ICD-9/BPA Code		80-90 Subtotal	91-95 Subtotal	1996	1997	1998	91-98 Subtotal	80-98 Total
CONGENITAL ANOMALIES OF URINARY SYSTEM (753)	**NUMBER**	**1019**	**592**	**113**	**114**	**119**	**938**	**1957**
	RATE	**2.15**	**2.91**	**3.00**	**3.10**	**3.15**	**2.97**	**2.48**
	Lower CI	**2.02**	**2.68**	**2.47**	**2.56**	**2.61**	**2.78**	**2.37**
	Upper CI	**2.29**	**3.16**	**3.60**	**3.72**	**3.77**	**3.17**	**2.59**
Renal Agenesis and Dysgenesis (7530)	NUMBER	191	94	12	14	12	132	323
	RATE	0.40	0.46	0.32	0.38	0.32	0.42	0.41
	Lower CI	0.35	0.37	0.16	0.21	0.16	0.35	0.37
	Upper CI	0.47	0.57	0.55	0.64	0.55	0.50	0.46
Cystic Kidney Disease (7531)	NUMBER	150	90	21	17	16	144	294
	RATE	0.32	0.44	0.56	0.46	0.42	0.46	0.37
	Lower CI	0.27	0.36	0.35	0.27	0.24	0.38	0.33
	Upper CI	0.37	0.54	0.85	0.74	0.69	0.54	0.42
Obstructive Defects of Renal Pelvis and Ureter (7532)	NUMBER	394	274	48	53	65	440	834
	RATE	0.83	1.35	1.27	1.44	1.72	1.39	1.06
	Lower CI	0.75	1.19	0.94	1.08	1.33	1.27	0.99
	Upper CI	0.92	1.52	1.69	1.89	2.19	1.53	1.13
Other Specified Anomalies of Kidney (7533)	NUMBER	84	55	9	7	8	79	163
	RATE	0.18	0.27	0.24	0.19	0.21	0.25	0.21
	Lower CI	0.14	0.20	0.11	0.08	0.09	0.20	0.18
	Upper CI	0.22	0.35	0.45	0.39	0.41	0.31	0.24
Other Specified Anomalies of Ureter (7534)	NUMBER	71	33	10	8	10	61	132
	RATE	0.15	0.16	0.27	0.22	0.26	0.19	0.17
	Lower CI	0.12	0.11	0.13	0.09	0.13	0.15	0.14
	Upper CI	0.19	0.23	0.49	0.42	0.48	0.25	0.20
Exstrophy of Urinary Bladder (7535)	NUMBER	17	10	0	2	1	13	30
	RATE	0.04	0.05	0	0.05	0.03	0.04	0.04
	Lower CI	0.02	0.02		0.01	0.00	0.02	0.03
	Upper CI	0.06	0.09		0.18	0.13	0.07	0.05

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Diagnostic Category and ICD-9/BPA Code		80-90 Subtotal	91-95 Subtotal	1996	1997	1998	91-98 Subtotal	80-98 Total
Atresia and Stenosis of Urethra And Bladder Neck (7536)	NUMBER	68	18	5	8	2	33	100
	RATE	0.14	0.09	0.13	0.22	0.05	0.10	0.13
	Lower CI	0.11	0.05	0.04	0.09	0.01	0.07	0.10
	Upper CI	0.18	0.14	0.30	0.42	0.18	0.15	0.15
Anomalies of Urachus (7537)	NUMBER	8	8	3	0	2	13	21
	RATE	0.02	0.04	0.08	0	0.05	0.04	0.03
	Lower CI	0.01	0.02	0.02		0.01	0.02	0.02
	Upper CI	0.03	0.08	0.23		0.18	0.07	0.04
Other Specified Anomalies of Bladder and Urethra (7538)	NUMBER	36	10	5	5	3	23	59
	RATE	0.08	0.05	0.13	0.14	0.08	0.07	0.07
	Lower CI	0.05	0.02	0.04	0.04	0.02	0.05	0.06
	Upper CI	0.11	0.09	0.30	0.31	0.22	0.11	0.10
CERTAIN CONGENITAL MUSCULOSKELETAL DEFORMITIES (754)	**NUMBER**	**3820**	**1356**	**191**	**165**	**163**	**1875**	**5695**
	RATE	**8.07**	**6.67**	**5.06**	**4.48**	**4.31**	**5.94**	**7.22**
	Lower CI	**7.81**	**6.32**	**4.37**	**3.83**	**3.68**	**5.68**	**7.03**
	Upper CI	**8.33**	**7.08**	**5.84**	**5.23**	**5.03**	**6.22**	**7.41**
Of Face, Skull and Jaw (7540)	NUMBER	57	27	4	3	3	37	94
	RATE	0.12	0.13	0.11	0.08	0.08	0.12	0.12
	Lower CI	0.09	0.09	0.03	0.02	0.02	0.08	0.10
	Upper CI	0.16	0.19	0.27	0.23	0.22	0.16	0.15
Of Sternocleidomastoid Muscle (7541)	NUMBER	14	14	0	0	0	14	28
	RATE	0.03	0.07	0	0	0	0.04	0.04
	Lower CI	0.02	0.04				0.02	0.02
	Upper CI	0.05	0.12				0.07	0.05
Of Spine (7542)	NUMBER	24	6	2	4	1	13	37
	RATE	0.05	0.03	0.05	0.11	0.03	0.04	0.05
	Lower CI	0.03	0.01	0.01	0.03	0.00	0.02	0.03
	Upper CI	0.08	0.06	0.18	0.27	0.13	0.07	0.06

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Diagnostic Category and ICD-9/BPA Code		80-90 Subtotal	91-95 Subtotal	1996	1997	1998	91-98 Subtotal	80-98 Total
Congenital Dislocation of Hip (7543)	NUMBER	1290	476	61	43	56	636	1926
	RATE	2.72	2.34	1.62	1.17	1.48	2.02	2.44
	Lower CI	2.58	2.14	1.24	0.85	1.12	1.86	2.33
	Upper CI	2.88	2.56	2.08	1.57	1.92	2.18	2.55
Genu Recurvatum and Bowing of Long Bones of Leg (7544)	NUMBER	8	12	2	4	1	19	27
	RATE	0.02	0.06	0.05	0.11	0.03	0.06	0.03
	Lower CI	0.01	0.03	0.01	0.03	0.00	0.04	0.02
	Upper CI	0.03	0.10	0.18	0.27	0.13	0.09	0.05
Varus Deformities of Feet (7545)	NUMBER	1489	505	78	69	67	719	2208
	RATE	3.14	2.48	2.07	1.88	1.77	2.28	2.80
	Lower CI	2.99	2.27	1.64	1.46	1.37	2.11	2.68
	Upper CI	3.31	2.71	2.58	2.37	2.25	2.45	2.92
Valgus Deformities of Feet (7546)	NUMBER	529	146	15	17	13	191	720
	RATE	1.12	0.72	0.40	0.46	0.34	0.61	0.91
	Lower CI	1.02	0.61	0.22	0.27	0.18	0.52	0.85
	Upper CI	1.22	0.84	0.65	0.74	0.59	0.70	0.98
Other Deformities of Feet (7547)	NUMBER	339	161	29	18	17	225	564
	RATE	0.72	0.79	0.77	0.49	0.45	0.71	0.71
	Lower CI	0.64	0.67	0.52	0.29	0.26	0.62	0.66
	Upper CI	0.80	0.92	1.10	0.77	0.72	0.81	0.78
Other Specified Musculoskeletal Deformities (7548)	NUMBER	70	10	0	7	5	22	92
	RATE	0.15	0.05	0	0.19	0.13	0.07	0.12
	Lower CI	0.12	0.02		0.08	0.04	0.04	0.09
	Upper CI	0.19	0.09		0.39	0.30	0.11	0.14
OTHER CONGENITAL ANOMALIES OF LIMBS (755)	**NUMBER**	**2208**	**910**	**136**	**142**	**156**	**1344**	**3552**
	RATE	**4.66**	**4.48**	**3.61**	**3.86**	**4.13**	**4.26**	**4.50**
	Lower CI	**4.47**	**4.19**	**3.03**	**3.25**	**3.51**	**4.03**	**4.35**
	Upper CI	**4.86**	**4.78**	**4.27**	**4.55**	**4.83**	**4.49**	**4.65**

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Diagnostic Category and ICD-9/BPA Code		80-90 Subtotal	91-95 Subtotal	1996	1997	1998	91-98 Subtotal	80-98 Total
Polydactyly (7550)	NUMBER	591	295	46	38	42	421	1012
	RATE	1.25	1.45	1.22	1.03	1.11	1.33	1.28
	Lower CI	1.15	1.29	0.89	0.73	0.80	1.21	1.20
	Upper CI	1.35	1.63	1.63	1.42	1.50	1.47	1.36
Syndactyly (7551)	NUMBER	430	200	37	29	38	304	734
	RATE	0.91	0.98	0.98	0.79	1.01	0.96	0.93
	Lower CI	0.82	0.85	0.69	0.53	0.71	0.86	0.86
	Upper CI	1.00	1.13	1.35	1.13	1.38	1.08	1.00
Reduction Deformities of Upper Limb (7552)	NUMBER	242	117	15	21	23	176	418
	RATE	0.51	0.58	0.40	0.57	0.61	0.56	0.53
	Lower CI	0.45	0.48	0.22	0.35	0.39	0.48	0.48
	Upper CI	0.58	0.69	0.65	0.87	0.91	0.65	0.58
Reduction Deformities of Lower Limb (7553)	NUMBER	113	57	12	2	12	83	196
	RATE	0.24	0.28	0.32	0.05	0.32	0.26	0.25
	Lower CI	0.20	0.21	0.16	0.01	0.16	0.21	0.21
	Upper CI	0.29	0.36	0.55	0.19	0.55	0.33	0.29
Other Anomalies of Upper Limb Including Shoulder Girdle (7555)	NUMBER	181	80	10	24	15	129	310
	RATE	0.38	0.39	0.27	0.65	0.40	0.41	0.39
	Lower CI	0.33	0.41	0.13	0.42	0.22	0.34	0.35
	Upper CI	0.44	0.39	0.49	0.97	0.65	0.39	0.44
Other Anomalies of Lower Limb Including Pelvic Girdle (7556)	NUMBER	601	135	13	25	25	198	799
	RATE	1.27	0.66	0.34	0.68	0.66	0.63	1.01
	Lower CI	1.17	0.56	0.18	0.44	0.43	0.54	0.94
	Upper CI	1.37	0.79	0.59	1.00	0.98	0.72	1.09
Other Specified Anomalies of Unspecified Limb (7558)	NUMBER	50	27	3	3	1	34	84
	RATE	0.11	0.13	0.08	0.08	0.03	0.11	0.11
	Lower CI	0.08	0.09	0.02	0.02	0.00	0.07	0.08
	Upper CI	0.14	0.19	0.23	0.23	0.13	0.15	0.13

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Diagnostic Category and ICD-9/BPA Code		80-90 Subtotal	91-95 Subtotal	1996	1997	1998	91-98 Subtotal	80-98 Total
OTHER CONGENITAL MUSCULOSKELETAL ANOMALIES (756)	**NUMBER**	**1091**	**513**	**97**	**129**	**89**	**828**	**1919**
	RATE	**2.30**	**2.52**	**2.57**	**3.51**	**2.35**	**2.62**	**2.43**
	Lower CI	**2.17**	**2.31**	**2.09**	**2.93**	**1.89**	**2.45**	**2.32**
	Upper CI	**2.44**	**2.75**	**3.14**	**4.16**	**2.90**	**2.81**	**2.54**
Anomalies of Skull and Face Bones (7560)	NUMBER	343	151	28	46	31	256	599
	RATE	0.72	0.74	0.74	1.25	0.82	0.81	0.76
	Lower CI	0.65	0.63	0.49	0.92	0.56	0.71	0.70
	Upper CI	0.81	0.87	1.07	1.67	1.16	0.92	0.82
Anomalies of Spine (7561)	NUMBER	109	67	14	18	6	105	214
	RATE	0.23	0.33	0.37	0.49	0.16	0.33	0.27
	Lower CI	0.19	0.26	0.20	0.29	0.06	0.27	0.24
	Upper CI	0.28	0.42	0.62	0.77	0.34	0.40	0.31
Other Anomalies of Ribs And Sternum (7563)	NUMBER	58	35	7	11	9	62	120
	RATE	0.12	0.17	0.19	0.30	0.24	0.20	0.15
	Lower CI	0.09	0.12	0.07	0.15	0.11	0.15	0.13
	Upper CI	0.16	0.24	0.38	0.53	0.45	0.25	0.18
Chondrodystrophy (7564)	NUMBER	56	22	5	3	6	36	92
	RATE	0.12	0.11	0.13	0.08	0.16	0.11	0.12
	Lower CI	0.09	0.07	0.04	0.02	0.06	0.08	0.09
	Upper CI	0.15	0.16	0.30	0.23	0.34	0.16	0.14
Osteodystrophies (7565)	NUMBER	35	16	1	1	3	21	56
	RATE	0.07	0.08	0.03	0.03	0.08	0.07	0.07
	Lower CI	0.05	0.05	0.00	0.00	0.02	0.04	0.05
	Upper CI	0.10	0.13	0.13	0.14	0.22	0.10	0.09
Anomalies of Diaphragm (7566)	NUMBER	182	61	16	9	14	100	282
	RATE	0.38	0.30	0.42	0.24	0.37	0.32	0.36
	Lower CI	0.33	0.23	0.24	0.11	0.20	0.26	0.32
	Upper CI	0.44	0.39	0.69	0.46	0.62	0.39	0.40

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Diagnostic Category and ICD-9/BPA Code		80-90 Subtotal	91-95 Subtotal	1996	1997	1998	91-98 Subtotal	80-98 Total
Anomalies of Abdominal Wall (7567)	NUMBER	182	71	11	27	18	127	309
	RATE	0.38	0.35	0.29	0.73	0.48	0.40	0.39
	Lower CI	0.33	0.27	0.15	0.48	0.28	0.34	0.35
	Upper CI	0.44	0.44	0.52	1.07	0.75	0.48	0.44
Other Specified Anomalies of Muscle, Tendon, Fascia, and Connective Tissue (7568)	NUMBER	126	90	15	14	1	120	246
	RATE	0.27	0.44	0.40	0.38	0.03	0.38	0.31
	Lower CI	0.22	0.36	0.22	0.21	0.00	0.32	0.27
	Upper CI	0.32	0.54	0.65	0.64	0.13	0.45	0.35
CONGENITAL ANOMALIES OF THE INTEGUMENT (757)	**NUMBER**	**657**	**326**	**31**	**41**	**33**	**431**	**1088**
	RATE	**1.39**	**1.60**	**0.82**	**1.11**	**0.87**	**1.37**	**1.38**
	Lower CI	**1.28**	**1.43**	**0.56**	**0.80**	**0.60**	**1.24**	**1.30**
	Upper CI	**1.50**	**1.79**	**1.17**	**1.51**	**1.23**	**1.50**	**1.46**
Hereditary Edema of the Legs (7570)	NUMBER	3	3	0	0	0	3	6
	RATE	0.01	0.01	0	0	0	0.01	0.01
	Lower CI	0.00	0.00				0.00	0.00
	Upper CI	0.02	0.04				0.03	0.02
Ichthyosis Congenita (7571)	NUMBER	19	7	1	1	0	9	28
	RATE	0.04	0.03	0.03	0.03	0	0.03	0.04
	Lower CI	0.02	0.01	0.00	0.00		0.01	0.02
	Upper CI	0.06	0.07	0.13	0.14		0.05	0.05
Dermatoglyphic Anomalies (7572)	NUMBER	325	180	14	23	11	228	553
	RATE	0.69	0.89	0.37	0.63	0.29	0.72	0.70
	Lower CI	0.61	0.76	0.20	0.40	0.15	0.63	0.64
	Upper CI	0.77	1.03	0.62	0.94	0.52	0.82	0.76
Other Specified Anomalies of Skin (7573)	NUMBER	195	85	9	10	17	121	316
	RATE	0.41	0.42	0.24	0.27	0.45	0.38	0.40
	Lower CI	0.36	0.33	0.11	0.13	0.26	0.32	0.36
	Upper CI	0.47	0.52	0.45	0.50	0.72	0.46	0.45

Number = Defects occurring in live births and stillbirths ≥ 20 weeks or ≥ 500 grams
CI = Approximate 95 per cent confidence intervals

Diagnostic Category and ICD-9/BPA Code		80-90 Subtotal	91-95 Subtotal	1996	1997	1998	91-98 Subtotal	80-98 Total
Specified Anomalies of Hair (7574)	NUMBER	3	5	0	0	0	5	8
	RATE	0.01	0.02	0	0	0	0.02	0.01
	Lower CI	0.00	0.01				0.01	0.00
	Upper CI	0.02	0.06				0.04	0.02
Specified Anomalies of Nails (7575)	NUMBER	26	10	0	0	3	13	39
	RATE	0.05	0.05	0	0	0.08	0.04	0.05
	Lower CI	0.04	0.02			0.02	0.02	0.04
	Upper CI	0.08	0.09			0.22	0.07	0.07
Specified Anomalies of Breast (7576)	NUMBER	73	30	7	7	2	46	119
	RATE	0.15	0.15	0.19	0.19	0.05	0.15	0.15
	Lower CI	0.12	0.10	0.07	0.08	0.01	0.11	0.12
	Upper CI	0.19	0.21	0.38	0.39	0.18	0.19	0.18
Other Specified Anomalies of the Integument (7579)	NUMBER	13	6	0	0	0	6	19
	RATE	0.03	0.03	0	0	0	0.02	0.02
	Lower CI	0.01	0.01				0.01	0.01
	Upper CI	0.05	0.06				0.04	0.04
CHROMOSOMAL ANOMALIES (758)	**NUMBER**	**688**	**384**	**58**	**74**	**87**	**603**	**1291**
	RATE	**1.45**	**1.89**	**1.54**	**2.01**	**2.30**	**1.91**	**1.64**
	Lower CI	**1.35**	**1.70**	**1.17**	**1.58**	**1.84**	**1.76**	**1.55**
	Upper CI	**1.57**	**2.09**	**1.99**	**2.52**	**2.84**	**2.07**	**1.73**
Down Syndrome (7580)	NUMBER	454	227	32	41	53	353	807
	RATE	0.96	1.12	0.85	1.11	1.40	1.12	1.02
	Lower CI	0.87	0.98	0.58	0.80	1.05	1.00	0.95
	Upper CI	1.05	1.27	1.20	1.51	1.83	1.24	1.10
Patau Syndrome (7581)	NUMBER	37	26	1	3	4	34	71
	RATE	0.08	0.13	0.03	0.08	0.11	0.11	0.09
	Lower CI	0.06	0.08	0.00	0.02	0.03	0.07	0.07
	Upper CI	0.11	0.19	0.13	0.23	0.27	0.15	0.11

Number = Defects occurring in live births and stillbirths ≥ 20 weeks or ≥ 500 grams
CI = Approximate 95 per cent confidence intervals

Diagnostic Category and ICD-9/BPA Code		80-90 Subtotal	91-95 Subtotal	1996	1997	1998	91-98 Subtotal	80-98 Total
Edwards Syndrome (7582)	NUMBER	80	51	9	13	8	81	161
	RATE	0.17	0.25	0.24	0.35	0.21	0.26	0.20
	Lower CI	0.13	0.19	0.11	0.19	0.09	0.20	0.17
	Upper CI	0.21	0.33	0.45	0.60	0.41	0.32	0.24
Autosomal Deletion Syndrome (7583)	NUMBER	22	12	3	4	3	22	44
	RATE	0.05	0.06	0.08	0.11	0.08	0.07	0.06
	Lower CI	0.03	0.03	0.02	0.03	0.02	0.04	0.04
	Upper CI	0.07	0.10	0.23	0.27	0.22	0.11	0.07
Other Conditions Due to Autosomal Anomalies (7585)	NUMBER	39	33	4	4	9	50	89
	RATE	0.08	0.16	0.11	0.11	0.24	0.16	0.11
	Lower CI	0.06	0.11	0.03	0.03	0.11	0.12	0.09
	Upper CI	0.11	0.23	0.27	0.27	0.45	0.21	0.14
Gonadal Dysgenesis (7586)	NUMBER	42	25	1	7	4	37	79
	RATE	0.09	0.12	0.03	0.19	0.11	0.12	0.10
	Lower CI	0.06	0.08	0.00	0.08	0.03	0.08	0.08
	Upper CI	0.12	0.18	0.13	0.39	0.27	0.16	0.12
Klinefelter Syndrome (7587)	NUMBER	7	6	4	1	1	12	19
	RATE	0.01	0.03	0.11	0.03	0.03	0.04	0.02
	Lower CI	0.01	0.01	0.03	0.00	0.00	0.02	0.01
	Upper CI	0.03	0.06	0.27	0.14	0.13	0.07	0.04
Other Conditions Due to Sex Chromosome Anomalies (7588)	NUMBER	7	4	1	1	5	11	18
	RATE	0.01	0.02	0.03	0.03	0.13	0.03	0.02
	Lower CI	0.01	0.01	0.00	0.00	0.04	0.02	0.01
	Upper CI	0.03	0.05	0.13	0.14	0.30	0.06	0.04
Conditions due to Anomalies of Unspecified Chromosomes (7589)	NUMBER	2	0	3	0	0	3	5
	RATE	0.00	0	0.08	0	0	0.01	0.01
	Lower CI	0.00		0.02			0.00	0.00
	Upper CI	0.01		0.23			0.03	0.01

Number = Defects occurring in live births and stillbirths ≥ 20 weeks or ≥ 500 grams
CI = Approximate 95 per cent confidence intervals

Diagnostic Category and ICD-9/BPA Code		80-90 Subtotal	91-95 Subtotal	1996	1997	1998	91-98 Subtotal	80-98 Total
OTHER AND UNSPECIFIED	**NUMBER**	**471**	**194**	**40**	**31**	**34**	**299**	**770**
CONGENITAL ANOMALIES (759)	**RATE**	**0.99**	**0.95**	**1.06**	**0.84**	**0.90**	**0.95**	**0.98**
	Lower CI	**0.91**	**0.82**	**0.76**	**0.57**	**0.62**	**0.84**	**0.91**
	Upper CI	**1.09**	**1.10**	**1.45**	**1.20**	**1.26**	**1.06**	**1.05**
Anomalies of Spleen (7590)	NUMBER	48	18	4	5	4	31	79
	RATE	0.10	0.09	0.11	0.14	0.11	0.10	0.10
	Lower CI	0.07	0.05	0.03	0.04	0.03	0.07	0.08
	Upper CI	0.13	0.14	0.27	0.31	0.27	0.14	0.12
Anomalies of Adrenal Gland (7591)	NUMBER	20	2	0	0	3	5	25
	RATE	0.04	0.01	0	0	0.08	0.02	0.03
	Lower CI	0.03	0.00			0.02	0.01	0.02
	Upper CI	0.07	0.03			0.22	0.04	0.05
Anomalies of Other Endocrine Glands (7592)	NUMBER	29	16	0	1	1	18	47
	RATE	0.06	0.08	0	0.03	0.03	0.06	0.06
	Lower CI	0.04	0.05		0.00	0.00	0.03	0.04
	Upper CI	0.09	0.13		0.14	0.13	0.09	0.08

Number = Defects occurring in live births and stillbirths ≥ 20 weeks or ≥ 500 grams
CI = Approximate 95 per cent confidence intervals

Diagnostic Category and ICD-9/BPA Code		80-90 Subtotal	91-95 Subtotal	1996	1997	1998	91-98 Subtotal	80-98 Total
Situs Inversus (7593)	NUMBER	31	18	10	5	5	38	69
	RATE	0.07	0.09	0.27	0.14	0.13	0.12	0.09
	Lower CI	0.04	0.05	0.13	0.04	0.04	0.09	0.07
	Upper CI	0.09	0.14	0.49	0.31	0.30	0.17	0.11
Conjoined Twins (7594)	NUMBER	10	1	0	0	1	2	12
	RATE	0.02	0.00	0	0	0.03	0.01	0.02
	Lower CI	0.01	0.00			0.00	0.00	0.01
	Upper CI	0.04	0.02			0.13	0.02	0.03
Tuberous Sclerosis (7595)	NUMBER	10	4	1	0	0	5	15
	RATE	0.02	0.02	0.03	0	0	0.02	0.02
	Lower CI	0.01	0.01	0.00			0.01	0.01
	Upper CI	0.04	0.05	0.13			0.04	0.03
Other Hamartoses, Not elsewhere Classified (7596)	NUMBER	10	1	1	0	1	3	13
	RATE	0.02	0.00	0.03	0	0.03	0.01	0.02
	Lower CI	0.01	0.00	0.00		0.00	0.00	0.01
	Upper CI	0.04	0.02	0.13		0.13	0.03	0.03
Mulitple Congenital Anomalies So Described (7597)	NUMBER	24	2	0	0	0	2	26
	RATE	0.05	0.01				0.01	0.03
	Lower CI	0.03	0.00				0.00	0.02
	Upper CI	0.08	0.03				0.02	0.05
Other Specified Anomalies (7598)	NUMBER	272	129	24	20	19	192	464
	RATE	0.57	0.63	0.64	0.54	0.50	0.61	0.59
	Lower CI	0.51	0.53	0.41	0.33	0.30	0.53	0.54
	Upper CI	0.65	0.75	0.95	0.84	0.78	0.70	0.64
Congenital Anomaly, Unspecified (7599)	NUMBER	17	3	0	0	0	3	20
	RATE	0.04	0.01	0	0	0	0.01	0.03
	Lower CI	0.02	0.00				0.00	0.02
	Upper CI	0.06	0.04				0.03	0.04

Number = Defects occurring in live births and stillbirths ≥ 20 weeks or ≥ 500 grams
CI = Approximate 95 per cent confidence intervals

Diagnostic Category and ICD-9/BPA Code		80-90 Subtotal	91-95 Subtotal	1996	1997	1998	91-98 Subtotal	80-98 Total
Situs Inversus (7593)	NUMBER	31	18	10	5	5	38	69
	RATE	0.07	0.09	0.27	0.14	0.13	0.12	0.09
	Lower CI	0.04	0.05	0.13	0.04	0.04	0.09	0.07
	Upper CI	0.09	0.14	0.49	0.31	0.30	0.17	0.11
Conjoined Twins (7594)	NUMBER	10	1	0	0	1	2	12
	RATE	0.02	0.00	0	0	0.03	0.01	0.02
	Lower CI	0.01	0.00			0.00	0.00	0.01
	Upper CI	0.04	0.02			0.13	0.02	0.03
Tuberous Sclerosis (7595)	NUMBER	10	4	1	0	0	5	15
	RATE	0.02	0.02	0.03	0	0	0.02	0.02
	Lower CI	0.01	0.01	0.00			0.01	0.01
	Upper CI	0.04	0.05	0.13			0.04	0.03
Other Hamartoses, Not elsewhere Classified (7596)	NUMBER	10	1	1	0	1	3	13
	RATE	0.02	0.00	0.03	0	0.03	0.01	0.02
	Lower CI	0.01	0.00	0.00		0.00	0.00	0.01
	Upper CI	0.04	0.02	0.13		0.13	0.03	0.03
Mulitple Congenital Anomalies So Described (7597)	NUMBER	24	2	0	0	0	2	26
	RATE	0.05	0.01				0.01	0.03
	Lower CI	0.03	0.00				0.00	0.02
	Upper CI	0.08	0.03				0.02	0.05
Other Specified Anomalies (7598)	NUMBER	272	129	24	20	19	192	464
	RATE	0.57	0.63	0.64	0.54	0.50	0.61	0.59
	Lower CI	0.51	0.53	0.41	0.33	0.30	0.53	0.54
	Upper CI	0.65	0.75	0.95	0.84	0.78	0.70	0.64
Congenital Anomaly, Unspecified (7599)	NUMBER	17	3	0	0	0	3	20
	RATE	0.04	0.01	0	0	0	0.01	0.03
	Lower CI	0.02	0.00				0.00	0.02
	Upper CI	0.06	0.04				0.03	0.04

Number = Defects occurring in live births and stillbirths ≥ 20 weeks or ≥ 500 grams
CI = Approximate 95 per cent confidence intervals

APPENDIX 3. SELECTED ANOMALIES AND RATES INCLUDING TERMINATIONS

Table A3.1 Selected Anomalies and Rates per 1,000 Including Terminations, 1997

Congenital Anomalies (CAs)	Number of Cases			Rates/1000 Total Births	
	Live	Still	Terminated	L + S	L + S + T
Anencephaly	4	5	12	0.24	0.57
Spina Bifida	12	2	0	0.38	0.38
Encephalocele	1	1	2	0.05	0.11
Hydrocephaly	13	4	5	0.46	0.60
Digestive System (*751..)	55	5	9	1.63	1.88
Urinary System (*753..)	100	14	9	3.10	3.34
Limb Anomalies (*755..)	132	10	8	3.86	4.08
Musculo-skeletal Anomalies (*756..)	118	11	7	3.51	3.70
Chromosome Anomalies (all)	62	12	32	2.01	2.88
Down Syndrome	40	1	15	1.11	1.52
Syndromes	18	2	2	0.54	0.60

*numbers indicate ICD-9/BPA code

Table A3.2 Selected Anomalies and Rates per 1,000 Including Terminations, 1998

Congenital Anomalies (CAs)	Number of Cases			Rates/1000 Total Births	
	Live	Still	Terminated	L + S	L + S + T
Anencephaly	2	2	8	0.11	0.32
Spina Bifida	9	4	1	0.34	0.37
Encephalocele	1	3	1	0.11	0.13
Hydrocephaly	8	0	4	0.21	0.32
Digestive System (*751..)	70	2	20	1.90	2.43
Urinary System (*753..)	112	7	22	3.15	3.73
Limb Anomalies (*755..)	143	13	39	4.13	5.16
Musculo-skeletal Anomalies (*756..)	77	12	13	2.35	2.70
Chromosome Anomalies (all)	74	13	37	2.30	3.28
Down Syndrome	50	3	19	1.40	1.90
Syndromes	19	0	2	0.50	0.56

*numbers indicate ICD-9/BPA code

APPENDIX 4. SELECTED ANOMALIES OUTSIDE ICD-9 CHAPTER XIV

Table A4 Alberta Congenital Anomalies Surveillance System Selected Disorders Outside Section XIV Anomaly Rates per 1,000 Total Births

Diagnostic Category and ICD-9/BPA Code		80-90 Subtotal	91-95 Subtotal	1996	1997	1998	91-98 Subtotal	80-98 Total
Neoplasms (140 – 239)	NUMBER	196	99	12	13	13	137	333
	RATE	0.41	0.49	0.32	0.35	0.34	0.43	0.42
	Lower CI	0.36	0.40	0.16	0.19	0.18	0.36	0.38
	Upper CI	0.48	0.59	0.55	0.60	0.59	0.51	0.47
Congenital Hypothyroidism (2439)	NUMBER	50	12	2	0	2	16	66
	RATE	0.11	0.06	0.05	0	0.05	0.05	0.08
	Lower CI	0.08	0.03	0.01		0.01	0.03	0.06
	Upper CI	0.14	0.10	0.18		0.18	0.08	0.11
Adrenogenital Disorders (2552)	NUMBER	25	7	0	1	1	9	34
	RATE	0.05	0.03	0	0.03	0.03	0.03	0.04
	Lower CI	0.03	0.01		0.00	0.00	0.01	0.03
	Upper CI	0.08	0.07		0.14	0.13	0.05	0.06
Amino Acid and Organic Acid Disorders (270)	NUMBER	60	16	2	0	2	20	80
	RATE	0.13	0.08	0.05	0	0.05	0.06	0.10
	Lower CI	0.10	0.05	0.01		0.01	0.04	0.08
	Upper CI	0.16	0.13	0.18		0.18	0.10	0.13
Disorders of CHO Transport and Metabolism (271)	NUMBER	33	18	0	1	2	21	54
	RATE	0.07	0.09	0	0.03	0.05	0.07	0.07
	Lower CI	0.05	0.05		0.00	0.01	0.04	0.05
	Upper CI	0.10	0.14		0.14	0.18	0.10	0.09

Number = Defects occurring in Live Births and Stillbirths ≥ 20 weeks or ≥ 500 g
CI = Approximate 95 per cent Confidence Intervals

Diagnostic Category and ICD-9/BPA Code		80-90 Subtotal	91-95 Subtotal	1996	1997	1998	91-98 Subtotal	80-98 Total
Disorders of Lipid Metabolism (272)	NUMBER	15	3	0	1	0	4	19
	RATE	0.03	0.01	0	0.03	0	0.01	0.02
	Lower CI	0.02	0.00		0.00		0.00	0.01
	Upper CI	0.05	0.04		0.14		0.03	0.04
Disorders of Mineral Metabolism (275)	NUMBER	25	7	3	0	2	12	37
	RATE	0.05	0.03	0.08	0	0.05	0.04	0.05
	Lower CI	0.03	0.01	0.02		0.01	0.02	0.03
	Upper CI	0.08	0.07	0.23		0.18	0.07	0.06
Cystic Fibrosis (27700)	NUMBER	36	18	1	1	1	21	57
	RATE	0.08	0.09	0.03	0.03	0.03	0.07	0.07
	Lower CI	0.05	0.05	0.00	0.00	0.00	0.04	0.05
	Upper CI	0.11	0.14	0.13	0.14	0.13	0.10	0.09
Mucopolysaccharidosis (2775)	NUMBER	4	1	0	0	0	1	5
	RATE	0.01	0.00	0	0	0	0.00	0.01
	Lower CI	0.00	0.00				0.00	0.00
	Upper CI	0.02	0.02				0.02	0.01
Hereditary Hemolytic Anemias (282)	NUMBER	56	7	4	2	1	14	70
	RATE	0.12	0.03	0.11	0.05	0.03	0.04	0.09
	Lower CI	0.09	0.01	0.03	0.01	0.00	0.02	0.07
	Upper CI	0.15	0.07	0.27	0.19	0.13	0.07	0.11
Coagulation Defects (2860-2864)	NUMBER	12	6	0	1	2	9	21
	RATE	0.03	0.03	0	0.03	0.05	0.03	0.03
	Lower CI	0.01	0.01		0.00	0.01	0.01	0.02
	Upper CI	0.04	0.06		0.14	0.18	0.05	0.04
Anterior Horn Cell Disease (335)	NUMBER	19	5	0	1	1	7	26
	RATE	0.04	0.02	0	0.03	0.03	0.02	0.03
	Lower CI	0.02	0.01		0.00	0.00	0.01	0.02
	Upper CI	0.06	0.06		0.14	0.13	0.05	0.05

Number = Defects occurring in Live Births and Stillbirths ≥ 20 weeks or ≥ 500 g
CI = Approximate 95 per cent Confidence Intervals

Diagnostic Category and ICD-9/BPA Code		80-90 Subtotal	91-95 Subtotal	1996	1997	1998	91-98 Subtotal	80-98 Total
Cerebral Cysts (3480)	NUMBER	29	16	4	1	2	23	52
	RATE	0.06	0.08	0.11	0.03	0.05	0.07	0.07
	Lower CI	0.04	0.05	0.03	0.00	0.01	0.05	0.05
	Upper CI	0.09	0.13	0.27	0.14	0.18	0.11	0.09
Muscular Dystrophies (3590, 3591, 3592 and 3598)	NUMBER	20	15	1	1	2	19	39
	RATE	0.04	0.07	0.03	0.03	0.05	0.06	0.05
	Lower CI	0.03	0.04	0.00	0.00	0.01	0.04	0.04
	Upper CI	0.07	0.12	0.13	0.14	0.18	0.09	0.07
Fetus or Newborn Affected by Maternal Ingestion of an Anticonvulsant (76070)	NUMBER	14	6	0	0	0	6	20
	RATE	0.03	0.03	0	0	0	0.02	0.03
	Lower CI	0.02	0.01				0.01	0.02
	Upper CI	0.05	0.06				0.04	0.04
Fetal Alcohol Syndrome/Effects (76076)	NUMBER	57	6	2	4	2	14	71
	RATE	0.12	0.03	0.05	0.11	0.05	0.04	0.09
	Lower CI	0.09	0.01	0.01	0.03	0.01	0.02	0.07
	Upper CI	0.16	0.06	0.18	0.27	0.18	0.07	0.11
Fetus or Newborn Affected by Maternal Ingestion of Other Drugs (76080)	NUMBER	3	0	0	0	0	0	3
	RATE	0.01	0	0	0	0	0	0.00
	Lower CI	0.00						0.00
	Upper CI	0.02						0.01

Number = Defects occurring in Live Births and Stillbirths ≥ 20 weeks or ≥ 500 g
CI = Approximate 95 per cent Confidence Intervals

APPENDIX 5. CONGENITAL ANOMALIES BY REGIONAL HEALTH AUTHORITY

Table A5. Congenital Anomaly Cases and Rate per 1,000 Total Live and Still Births, by Regional Health Authority, 1996-1998

Region	1996		1997		1998	
	Cases	Rate (95% CI)	Cases	Rate (95% CI)	Cases	Rate (95% CI)
ALBERTA	**1196**	**31.71 (29.94-33.57)**	**1117**	**30.35 (28.60-32.19)**	**1183**	**31.22 (29.47-33.06)**
1. Chinook	89	41.13 (33.05-50.60)	92	40.85 (32.95-50.09)	73	35.08 (27.51-44.09)
2. Palliser	71	60.74 (47.46-76.57)	68	57.05 (44.33-72.28)	53	42.43 (31.81-55.46)
3. Headwaters	33	38.51 (26.54-54.09)	29	31.76 (21.30-45.62)	25	27.81 (18.02-41.05)
4. Calgary	388	34.57 (31.22-38.19)	369	33.15 (29.86-36.71)	487	41.75 (38.13-45.63)
5. Health Authority 5	17	26.15 (15.26-41.82)	14	22.36 (12.25-37.43)	22	34.87 (21.88-52.76)
6. David Thompson	77	29.60 (23.38-36.99)	67	26.61 (20.63-33.77)	67	26.11 (20.25-33.14)
7. East Central	21	19.72 (12.22-30.12)	30	29.38 (19.85-41.95)	29	27.72 (18.59-39.82)
8. WestView	28	24.20 (16.10-34.98)	34	29.26 (20.29-40.90)	25	21.82 (14.14-32.20)
9. Crossroads	37	37.49 (26.42-51.69)	35	37.27 (25.99-51.86)	14	24.78 (13.57-41.48)
10. Capital	260	26.52 (23.40-29.96)	245	26.54 (23.32-30.09)	262	26.19 (23.12-29.56)
11. Aspen	21	17.54 (10.88-26.80)	24	21.70 (13.92-32.28)	27	23.91 (15.78-34.80)
12. Lakeland	62	40.90 (31.38-52.40)	56	37.23 (28.15-48.32)	42	27.47 (19.82-37.15)
13. Mistahia	39	29.28 (20.84-40.05)	13	10.13 (5.40-17.28)	17	12.78 (7.46-20.44)
14. Peace	6	18.02 (6.60-38.74)	7	23.57 (9.48-48.10)	5	15.58 (5.04-35.77)
15. Keeweetinok	16	30.08 (17.22-48.76)	10	19.72 (9.47-36.09)	14	26.52 (14.52-44.38)
16. Northern Lights	23	38.21 (24.25-57.31)	20	34.90 (21.35-53.87)	12	20.24 (10.47-35.23)
17. Northwestern	8	15.30 (6.61-29.91)	4	7.39 (2.00-18.52)	9	17.51 (8.02-18.52)

APPENDIX 6. ANNUAL SELECTED ANOMALIES FOR REGIONAL HEALTH AUTHORITIES

Table A6.1 Cleft lip ± cleft palate: anomaly count and rates per 1000 total births,‡ 1994 – 1998.

	1994 (95% CI)	1995 (95% CI)	1996 (95% CI)	1997 (95% CI)	1998† (95% CI)
ALBERTA	**38 0.96 (0.68, 1.31)**	**52 1.34 (1.00, 1.76)**	**45 1.19 (0.87, 1.60)**	**33 0.90 (0.62, 1.26)**	**47 1.24 (0.91, 1.65)**
1. Chinook	0	4 1.73 (0.47, 4.33)	5 2.31 (0.75, 5.30)	1 0.44 (0.02, 2.25)	5 2.40 (0.78, 5.52)
2. Palliser	2 1.75 (0.20, 6.03)	1 0.78 (0.03, 3.95)	0	0	1 0.80 (0.03, 4.05)
3. Headwaters	2 2.23 (0.25, 7.65)	1 1.19 (0.05, 6.02)	1 1.17 (0.05, 5.90)	0	0
4. Calgary	8 0.69 (0.30, 1.35)	14 1.24 (0.68, 2.07)	15 1.34 (0.75, 2.20)	6 0.54 (0.20, 1.16)	10 0.86 (0.41, 1.57)
5. Health Authority 5	1 1.55 (0.06, 7.84)	2 2.98 (0.34,10.24)	1 1.54 (0.06, 7.78)	0	0
6. David Thompson	4 1.52 (0.41, 3.80)	4 1.52 (0.41, 3.81)	3 1.15 (0.23, 3.27)	4 1.59 (0.43, 3.98)	9 3.51 (1.61, 6.62)
7. East Central	1 0.83 (0.03, 4.22)	0	0	1 0.98 (0.04, 4.96)	2 1.91 (0.22, 6.57)
8. WestView	2 1.77 (0.20, 6.07)	0	1 0.86 (0.03, 4.37)	3 2.58 (0.52, 7.32)	2 1.75 (0.20, 6.00)
9. Crossroads	0	1 0.91 (0.04, 4.63)	0	1 1.06 (0.04, 5.38)	0
10. Capital	9 0.84 (0.39, 1.59)	11 1.08 (0.54, 1.93)	11 1.12 (0.56, 2.00)	8 0.87 (0.37, 1.69)	11 1.10 (0.55, 1.96)
11. Aspen	1 0.76 (0.03, 3.82)	1 0.82 (0.03, 4.16)	0	2 1.81 (0.21, 6.21)	0
12. Lakeland	3 1.83 (0.37, 5.19)	4 2.53 (0.68, 6.35)	0	3 1.99 (0.40, 5.65)	1 0.65 (0.03, 3.31)
13. Mistahia	4 3.02 (0.82, 7.57)	4 2.97 (0.80, 7.44)	4 3.00 (0.81, 7.52)	1 0.78 (0.03, 3.94)	1 0.75 (0.03, 3.80)
14. Peace	1 3.03 (0.12,15.33)	0	0	0	1 3.12 (0.12, 15.76)
15. Keeweetinok	0	3 5.48 (1.11, 15.53)	1 1.88 (0.08, 9.51)	2 3.94 (0.45,13.55)	3 5.68 (1.15, 16.09)
16. Northern Lights	0	1 1.73 (0.07, 8.75)	1 1.66 (0.07, 8.41)	1 1.75 (0.07, 8.83)	1 1.69 (0.07, 8.53)
17. Northwestern	0	1 1.78 (0.07, 9.02)	2 3.82 (0.44, 13.14)	0	0

* 1st number reported represents number of defects; 2nd number represents the rate

† N.B. Boundary changes in 1998 ‡ Total births = Live births + stillbirths

Table A6.2 Cleft palate: anomaly count and rates per 1000 total births,‡ 1994 – 1998.

	1994 (95% CI)	1995 (95% CI)	1996 (95% CI)	1997 (95% CI)	1998† (95% CI)
ALBERTA	**28 0.70 (0.47, 1.02)**	**34 0.90 (0.63, 1.26)**	**23 0.61 (0.39, 0.91)**	**38 1.03 (0.73, 1.42)**	**28 0.74 (0.49, 1.07)**
1. Chinook	0	1 0.43 (0.02, 2.19)	1 0.46 (0.02, 2.34)	7 3.11 (1.25, 6.34)	0
2. Palliser	0	0	0	1 0.84 (0.03, 4.24)	0
3. Headwaters	1 1.11 (0.04, 5.63)	0	0	1 1.10 (0.04, 5.54)	1 1.11 (0.04, 5.63)
4. Calgary	9 0.78 (0.36, 1.46)	10 0.88 (0.42, 1.61)	5 0.45 (0.14, 1.02)	5 0.45 (0.15, 1.03)	10 0.86 (0.41, 1.57)
5. Health Authority 5	2 3.10 (0.35, 10.65)	0	0	0	0
6. David Thompson	4 1.52 (0.41, 3.80)	3 1.14 (0.23, 3.23)	3 1.15 (0.23, 3.27)	6 2.38 (0.87, 5.12)	3 1.17 (0.24, 3.31)
7. East Central	0	0	1 0.94 (0.04, 4.75)	2 1.96 (0.22, 6.73)	1 0.96 (0.04, 4.84)
8. WestView	1 0.88 (0.04, 4.47)	2 1.62 (0.19, 5.57)	2 1.73 (0.20, 5.94)	0	0
9. Crossroads	0	1 0.91 (0.04, 4.63)	0	3 3.19 (0.64, 9.03)	0
10. Capital	8 0.75 (0.32, 1.47)	11 1.08 (0.54, 1.93)	7 0.71 (0.29, 1.46)	10 1.08 (0.52, 1.98)	8 0.80 (0.35, 1.56)
11. Aspen	1 0.76 (0.03, 3.82)	1 0.82 (0.03, 4.16)	0	1 0.90 (0.04, 4.58)	1 0.89 (0.04, 4.48)
12. Lakeland	0	2 1.27 (0.14, 4.35)	1 0.66 (0.03, 3.34)	1 0.66 (0.03, 3.36)	4 2.62 (0.71, 6.55)
13. Mistahia	2 1.51 (0.17, 5.19)	2 1.49 (0.17, 5.11)	1 0.75 (0.03, 3.80)	0	0
14. Peace	0	0	0	0	0
15. Keeweetinok	0	0	1 1.88 (0.08, 9.51)	0	0
16. Northern Lights	0	1 1.73 (0.07, 8.75)	1 1.66 (0.07, 8.41)	0	0
17. Northwestern	0	0	0	1 1.85 (0.07, 9.35)	0

* 1st number reported represents number of defects; 2nd number represents the rate

† N.B. Boundary changes in 1998 ‡ Total births = Live births + stillbirths

Table A6.3 Down Syndrome: anomaly count and rates per 1000 total births,‡ 1994 – 1998.

	1994 (95% CI)	1995 (95% CI)	1996 (95% CI)	1997 (95% CI)	1998† (95% CI)
ALBERTA	**44 1.11 (0.81, 1.49)**	**49 1.26 (0.94, 1.67)**	**32 0.85 (0.58, 1.20)**	**41 1.11 (0.80, 1.51)**	**53 1.40 (1.05, 1.83)**
1. Chinook	1 0.43 (0.02, 2.19)	3 1.30 (0.26, 3.67)	1 0.46 (0.02, 2.34)	3 1.33 (0.27, 3.77)	3 1.44 (0.29, 4.08)
2. Palliser	3 2.63 (0.53, 7.45)	4 3.12 (0.84, 7.82)	2 1.71 (0.20, 5.88)	0	4 3.20 (0.86, 8.02)
3. Headwaters	1 1.11 (0.04, 5.63)	5 5.95 (1.93, 13.67)	0	3 3.29 (0.66, 9.30)	2 2.22 (0.25, 7.64)
4. Calgary	8 0.69 (0.30, 1.35)	20 1.76 (1.08, 2.72)	13 1.16 (0.62, 1.98)	11 0.99 (0.49, 1.76)	16 1.37 (0.79, 2.22)
5. Health Authority 5	1 1.55 (0.06, 7.84)	0	0	2 3.19 (0.36,10.98)	2 3.17 (0.36, 10.89)
6. David Thompson	3 1.14 (0.23, 3.22)	0	4 1.54 (0.42, 3.85)	3 1.19 (0.24, 3.37)	4 1.56 (0.42, 3.90)
7. East Central	0	1 0.92 (0.04, 4.65)	0	2 1.96 (0.22, 6.73)	0
8. WestView	1 0.88 (0.04, 4.47)	0	1 0.86 (0.03, 4.37)	1 0.86 (0.03, 4.36)	1 0.87 (0.03, 4.42)
9. Crossroads	2 1.76 (0.20, 6.03)	0	0	3 3.19 (0.64, 9.03)	2 3.54 (0.40, 12.16)
10. Capital	15 1.41 (0.79, 2.31)	9 0.99 (0.47, 1.80)	6 0.61 (0.22, 1.32)	8 0.87 (0.37, 1.69)	16 1.60 (0.92, 2.59)
11. Aspen	3 2.27 (0.46, 6.42)	0	0	3 2.71 (0.55, 7.68)	2 1.77 (0.20, 6.09)
12. Lakeland	3 1.83 (0.37, 5.19)	3 1.90 (0.38, 5.38)	1 0.66 (0.03, 3.34)	1 0.66 (0.03, 3.36)	1 0.65 (0.03, 3.31)
13. Mistahia	0	2 1.49 (0.17, 5.11)	2 1.50 (0.17, 5.16)	1 0.78 (0.03, 3.94)	0
14. Peace	1 3.03 (0.12,15.33)	1 3.02 (0.12,15.29)	0	0	0
15. Keeweetinok	1 1.67 (0.07, 8.46)	1 1.83 (0.07, 9.25)	2 3.76 (0.43, 12.92)	0	0
16. Northern Lights	0	0	0	0	0
17. Northwestern	1 1.88 (0.08, 9.51)	0	0	0	0

* 1st number reported represents number of defects; 2nd number represents the rate
† N.B. Boundary changes in 1998 ‡ Total births = Live births + stillbirths

Table A6.4 Neural tube defects: anomaly count and rates per 1000 total births,‡ 1994 – 1998.

	1994 (95% CI)	1995 (95% CI)	1996 (95% CI)	1997 (95% CI)	1998† (95% CI)
ALBERTA	**32 0.81 (0.55. 1.14)**	**39 1.01 (0.72, 1.38)**	**24 0.64 (0.41, 0.95)**	**25 0.68 (0.44, 1.00)**	**21 0.56 (0.34, 0.85)**
1. Chinook	0	2 0.87 (0.10, 2.97)	3 1.39 (0.28, 3.92)	1 0.44 (0.02, 2.25)	0
2. Palliser	0	2 1.56 (0.18, 5.36)	3 2.57 (0.52, 7.27)	1 0.84 (0.03, 4.24)	1 0.80 (0.03, 4.05)
3. Headwaters	0	0	0	1 1.10 (0.04, 5.54)	0
4. Calgary	9 0.78 (0.36, 1.46)	6 0.53 (0.19, 1.14)	6 0.53 (0.20, 1.15)	5 0.45 (0.15, 1.03)	11 0.94 (0.47, 1.68)
5. Health Authority 5	1 1.55 (0.06, 7.84)	0	0	0	1 1.58 (0.06, 8.02)
6. David Thompson	2 0.76 (0.09, 2.60)	3 1.14 (0.23, 3.23)	4 1.54 (0.42, 3.85)	0	0
7. East Central	0	2 1.84 (0.21, 6.31)	1 0.94 (0.04, 4.75)	0	1 0.96 (0.04, 4.84)
8. WestView	1 0.88 (0.04, 4.47)	1 0.81 (0.03, 4.10)	0	4 3.45 (0.93, 8.63)	0
9. Crossroads	0	0	3 3.03 (0.61, 8.59)	1 1.06 (0.04, 5.38)	0
10. Capital	14 1.31 (0.72, 2.20)	11 1.08 (0.54, 1.93)	1 0.10 (0.00, 0.52)	6 0.65 (0.24, 1.40)	6 0.60 (0.22, 1.29)
11. Aspen	1 0.76 (0.03, 3.82)	3 2.47 (0.50, 6.99)	0	2 1.81 (0.21, 6.21)	0
12. Lakeland	1 0.61 (0.02, 3.09)	4 2.53 (0.68, 6.35)	2 1.32 (0.15, 4.53)	2 1.33 (0.15, 4.57)	1 0.65 (0.03, 3.31)
13. Mistahia	1 0.76 (0.03, 3.82)	1 0.74 (0.03, 3.76)	0	0	0
14. Peace	0	0	0	0	0
15. Keeweetinok	2 3.34 (0.38, 11.49)	1 1.83 (0.07, 9.25)	1 1.88 (0.08, 9.51)	1 1.97 (0.08, 9.98)	0
16. Northern Lights	0	2 3.46 (0.39, 11.89)	0	1 1.75 (0.07, 8.83)	0
17. Northwestern	0	1 1.78 (0.07, 9.02)	0	0	0

* 1st number reported represents number of defects; 2nd number represents the rate

† N.B. Boundary changes in 1998 ‡ Total births = Live births + stillbirths

Table A6.5 Cardiac - Bulbis cordis anomalies and defects of septal closure (745..): anomaly count and rates per 1000 total births,‡ 1994 – 1998.

	1994 (95% CI)	1995 (95% CI)	1996 (95% CI)	1997 (95% CI)	1998† (95% CI)
ALBERTA	**222 5.59 (4.88, 6.37)**	**201 5.18 (4.49, 5.95)**	**232 6.15 (5.39, 7.00)**	**212 5.76 (5.01, 6.59)**	**197 5.21 (4.51, 5.99)**
1. Chinook	14 5.99 (3.28, 10.02)	20 8.65 (5.29, 13.35)	20 9.24 (5.65, 14.26)	34 15.10 (10.47, 21.11)	21 10.09 (6.26, 15.42)
2. Palliser	8 7.02 (3.03, 13.72)	4 3.12 (0.84, 7.82)	15 12.83 (7.19, 21.12)	11 9.23 (4.61, 16.44)	9 7.21 (3.30, 13.59)
3. Headwaters	1 1.11 (0.04, 5.63)	8 9.52 (4.11, 18.62)	5 5.83 (1.89, 13.40)	8 8.76 (3.79, 17.13)	4 4.45 (1.20, 11.15)
4. Calgary	78 6.72 (5.32, 8,39)	55 4.85 (3.66, 6.31)	77 6.86 (5.42, 8.57)	56 5.03 (3.80, 6.53)	54 4.63 (3.48, 6.04)
5. Health Authority 5	6 9.30 (3.41, 20.00)	6 8.94 (3.28, 19.23)	5 7.69 (2.49, 17.67)	3 4.79 (0.97, 13.57)	3 4.75 (0.96, 13.46)
6. David Thompson	12 4.55 (2.35, 7.92)	9 3.42 (1.56, 6.45)	8 3.08 (1.33, 6.01)	10 3.97 (1.91, 7.27)	13 5.07 (2.70, 8.64)
7. East Central	3 2.50 (0.50, 7.08)	5 4.59 (1.49, 10.54)	4 3.76 (1.01, 9.41)	1 0.98 (0.04, 4.96)	6 5.74 (2.10, 12.33)
8. WestView	3 2.65 (0.53, 7.50)	4 3.24 (0.88, 8.13)	6 5.19 (1.90, 11.15)	8 6.89 (2.98, 13.47)	5 4.36 (1.41, 10.02)
9. Crossroads	4 3.51 (0.95, 8.80)	5 4.57 (1.48, 10.50)	3 3.03 (0.61, 8.59)	9 9.56 (4.38, 18.04)	6 10.62 (3.89, 22.83)
10. Capital	56 5.25 (3.97, 6.81)	50 4.93 (3.66, 6.49)	50 5.10 (3.79, 6.73)	38 4.12 (2.92, 5.65)	42 4.20 (3.03, 5.68)
11. Aspen	8 6.04 (2.61, 11.82)	2 1.64 (0.19, 5.65)	3 2.51 (0.51, 7.10)	8 7.23 (3.13, 14.14)	10 8.86 (4.25, 16.21)
12. Lakeland	12 7.33 (3.80, 12.77)	9 5.70 (2.61, 10.75)	17 11.21 (6.54, 17.93)	15 9.97 (5.59, 16.42)	11 7.19 (3.60, 12.82)
13. Mistahia	9 6.80 (3.11, 12.83)	13 9.66 (5.15, 16.47)	10 7.51 (3.61, 13.74)	7 5.46 (2.19, 11.13)	1 0.75 (0.03, 3.80)
14. Peace	2 6.06 (0.69, 20.82)	5 15.11 (4.89, 34.69)	0	0	0
15. Keeweetinok	3 5.02 (1.01, 14.21)	1 1.83 (0.07, 9.25)	3 5.64 (1.14, 15.97)	0	4 7.58 (2.05, 18.98)
16. Northern Lights	1 1.72 (0.07, 8.71)	3 5.19 (1.05, 14.70)	4 6.64 (1.79, 16.64)	3 5.24 (1.06, 14.83)	2 3.37 (0.38, 11.59)
17. Northwestern	2 3.76 (0.43, 12.92)	2 3.57 (0.41, 12.25)	2 3.82 (0.44, 13.14)	1 1.85 (0.07, 9.35)	6 11.67 (4.28, 25.10)

* 1st number reported represents number of defects; 2nd number represents the rate
† N.B. Boundary changes in 1998 ‡ Total births = Live births + stillbirths

Table A6.6 Cardiac - other anomalies of heart, mainly valvular defects (746..): anomaly count and rates per 1000 total births,‡ 1994 – 1998.

	1994 (95% CI)	1995 (95% CI)	1996 (95% CI)	1997 (95% CI)	1998† (95% CI)
ALBERTA	**90 2.27 (1.82, 2.78)**	**88 2.24 (1.80, 2.77)**	**118 3.13 (2.59, 3.75)**	**113 3.13 (2.58, 3.75)**	**113 2.99 (2.46, 3.59)**
1. Chinook	5 2.14 (0.69, 4.91)	9 3.89 (1.78, 7.34)	9 4.16 (1.90, 7.84)	13 5.77 (3.08, 9.84)	13 6.25 (3.33, 10.65)
2. Palliser	3 2.63 (0.53, 7.45)	1 0.78 (0.03, 3.95)	3 2.57 (0.52, 7.27)	3 2.52 (0.51, 7.13)	3 2.40 (0.48, 6.80)
3. Headwaters	0	1 1.19 (0.05, 6.02)	1 1.17 (0.05, 5.90)	4 4.38 (1.18, 10.97)	0
4. Calgary	22 1.90 (1.19, 2.87)	18 1.59 (0.94, 2.51)	37 3.30 (2.32, 4.55)	30 2.69 (1.82, 3.85)	25 2.14 (1.39, 3.16)
5. Health Authority 5	1 1.55 (0.06, 7.84)	2 2.98 (0.34, 10.24)	0	1 1.60 (0.06, 8.08)	2 3.17 (0.36, 10.89)
6. David Thompson	4 1.52 (0.41, 3.80)	10 3.80 (1.82, 6.95)	7 2.69 (1.08, 5.49)	6 2.38 (0.87, 5.12)	11 4.29 (2.14, 7.64)
7. East Central	1 0.83 (0.03, 4.22)	5 4.59 (1.49, 10.54)	2 1.88 (0.21, 6.45)	0	4 3.82 (1.03, 9.58)
8. WestView	3 2.65 (0.53, 7.50)	1 0.81 (0.03, 4.10)	4 3.46 (0.93, 8.66)	4 3.45 (0.93, 8.63)	7 6.11 (2.46, 12.47)
9. Crossroads	6 5.27 (1.93, 11.33)	4 3.66 (0.99, 9.16)	1 1.01 (0.04, 5.12)	10 10.63 (5.10, 19.45)	3 5.31 (1.07, 15.04)
10. Capital	26 2.44 (1.59, 3.57)	21 2.07 (1.28, 3.16)	30 3.06 (2.07, 4.37)	32 3.47 (2.37, 4.90)	24 2.40 (1.54, 3.57)
11. Aspen	1 0.76 (0.03, 3.82)	3 2.47 (0.50, 6.99)	0	0	4 3.54 (0.96, 8.88)
12. Lakeland	10 6.11 (2.94, 11.18)	5 3.17 (1.02, 7.27)	10 6.60 (3.17, 12.07)	4 2.66 (0.72, 6.66)	6 3.92 (1.44, 8.44)
13. Mistahia	5 3.78 (1.22, 8.68)	3 2.23 (0.45, 6.31)	2 1.50 (0.17, 5.16)	2 1.56 (0.18, 5.36)	0
14. Peace	0	1 3.02 (0.12, 15.29)	2 6.01 (0.69, 20.64)	1 3.37 (0.13, 17.04)	1 3.12 (0.12, 15.76)
15. Keeweetinok	2 3.34 (0.38, 11.49)	2 3.66 (0.42, 12.56)	2 3.76 (0.43, 12.92)	1 1.97 (0.08, 9.98)	3 5.68 (1.15, 16.09)
16. Northern Lights	0	2 3.46 (0.39, 11.89)	3 4.98 (1.01, 14.11)	2 3.49 (0.40, 11.99)	1 1.69 (0.07, 8.53)
17. Northwestern	1 1.88 (0.08, 9.51)	0	5 9.56 (3.09, 21.96)	0	6 11.67 (4.28, 25.10)

* 1st number reported represents number of defects; 2nd number represents the rate

† N.B. Boundary changes in 1998 ‡ Total births = Live births + stillbirths

Table A6.7 Cardiac (745.. and 746..): anomaly count and rates per 1000 total births,‡ 1994 – 1998.

	1994 (95% CI)	1995 (95% CI)	1996 (95% CI)	1997 (95% CI)	1998† (95% CI)
ALBERTA	**312 7.85 (7.01, 8.78)**	**289 7.45 (6.62, 8.36)**	**350 9.28 (8.34, 10.31)**	**327 8.89 (7.95, 9.90)**	**310 8.20 (7.31, 9.16)**
1. Chinook	19 8.12 (4.90, 12.67)	29 12.54 (8.41, 18.02)	29 13.39 (8.98, 19.24)	47 20.87 (15.35, 27.77)	34 16.34 (11.33, 22.84)
2. Palliser	11 9.65 (4.82, 17.19)	5 3.90 (1.26, 8.96)	18 15.40 (9.14, 24.31)	14 11.74 (6.43, 19.66)	12 9.61 (4.97, 16.73)
3. Headwaters	1 1.11 (0.04, 5.63)	9 10.71 (4.90, 20.21)	6 7.00 (2.57, 15.05)	12 13.14 (6.80, 22.88)	4 4.45 (1.20, 11.15)
4. Calgary	100 8.62 (7.02, 10.48)	73 6.44 (5.05, 8.09)	114 10.16 (8.38, 12.21)	86 7.73 (6.18, 9.55)	79 6.77 (5.37, 8.44)
5. Health Authority 5	7 10.85 (4.36, 22.15)	8 11.92 (5.15, 23.31)	5 7.69 (2.49, 17.67)	4 6.39 (1.73, 16.01)	5 7.92 (2.56, 18.20)
6. David Thompson	16 6.07 (3.47, 9.83)	19 7.22 (4.35, 11.26)	15 5.77 (3.23, 9.49)	16 6.35 (3.64, 10.30)	24 9.35 (6.00, 13.91)
7. East Central	4 3.33 (0.90, 8.35)	10 9.18 (4.41, 16.80)	6 5.63 (2.06, 12.11)	1 0.98 (0.04, 4.96)	10 9.56 (4.59, 17.79)
8. WestView	6 5.30 (1.94, 11.39)	5 4.06 (1.31, 9.31)	10 8.64 (4.15, 15.82)	12 10.34 (5.35, 17.99)	12 10.47 (5.42, 18.23)
9. Crossroads	10 8.78 (4.22, 16.07)	9 8.23 (3.77, 15.62)	4 4.04 (1.09, 10.13)	19 20.19 (12.17, 31.50)	9 15.93 (7.29, 30.05)
10. Capital	82 7.69 (6.12, 9.54)	71 7.00 (5.47, 8.82)	80 8.16 (6.48, 10.17)	70 7.58 (5.92, 9.59)	66 6.60 (5.11, 8.39)
11. Aspen	9 6.80 (3.11, 12.82)	5 4.11 (1.33, 9.44)	3 2.51 (0.51, 7.10)	8 7.23 (3.13, 14.14)	14 12.40 (6.79, 20.76)
12. Lakeland	22 13.45 (8.44, 20.35)	14 8.87 (4.86, 14.84)	27 17.81 (11.75, 25.91)	19 12.63 (7.62, 19.71)	17 11.12 (6.49, 17.78)
13. Mistahia	14 10.58 (5.79, 17.71)	16 11.89 (6.81, 19.27)	12 9.01 (4.66, 15.68)	9 7.01 (3.21, 13.23)	1 0.75 (0.03, 3.80)
14. Peace	2 6.06 (0.69, 20.82)	6 18.13 (6.64, 38.98)	2 6.01 (0.69, 20.64)	1 3.37 (0.13, 17.04)	1 3.12 (0.12, 15.76)
15. Keeweetinok	5 8.36 (2.70, 19.20)	3 5.48 (1.11, 15.53)	5 9.40 (3.04, 21.58)	1 1.97 (0.08, 9.98)	7 13.26 (5.33, 27.06)
16. Northern Lights	1 1.72 (0.07, 8.71)	5 8.65 (2.80, 19.87)	7 11.63 (4.67, 22.73)	5 8.73 (2.82, 20.04)	3 5.06 (1.02, 14.33)
17. Northwestern	3 5.64 (1.14, 15.97)	2 3.57 (0.41, 12.25)	7 13.38 (5.38, 27.31)	1 1.85 (0.07, 9.35)	12 23.35 (12.08, 40.64)

* 1st number reported represents number of defects; 2nd number represents the rate

† N.B. Boundary changes in 1998 ‡ Total births = Live births + stillbirths

APPENDIX 7. SELECTED ANOMALY RATES FOR ALBERTA AND OTHER JURISDICTIONS REPORTING TO ICBDMS*, 1997

Table A7.1 Rates of selected congenital anomalies per 1,000 total ‡ births with 95 per cent confidence intervals.

Anomaly	Alberta L,S	Alberta L,S,T	Atlanta L,S	Atlanta L,S,T	Australia L,S	Australia L,S,T	Central-East-France L,S	Central-East-France L,S,T
Anencephaly	**0.24** (0.11,0.46)	**0.57** (0.35,0.87)	**0.09** (0.02,0.23)	**0.18** (0.08,0.36)	**0.16** (0.11,0.21)	**0.43** (0.35,0.52)	**0**	**0.13** (0.07,0.22)
Spina Bifida	**0.38** (0.21,0.64)	0 termin.	**0.30** (0.16,0.51)	**0.37** (0.21,0.60)	**0.29** (0.23,0.37)	**0.55** (0.46,0.65)	**0.11** (0.05,0.19)	**0.40** (0.29,0.55)
Encephalocele	**0.05** (0.01,0.19)	**0.11** (0.03,0.27)	**0.09** (0.02,0.23)	**0.12** (0.04,0.27)	**0.05** (0.02,0.08)	**0.11** (0.07,0.16)	**0.01** (0.00,0.05)	**0.13** (0.07,0.22)
All NTDs	**0.68** (0.44,1.00)	**1.06** (0.75,1.45)	**0.49** (0.30,0.74)	**0.67** (0.45,0.96)	**0.50** (0.42,0.59)	**1.09** (0.97,1.23)	**0.12** (0.06,0.21)	**0.66** (0.51,0.83)
Hydrocephaly	**0.46** (0.27,0.74)	**0.60** (0.38,0.90)	**0.32** (0.18,0.54)	**0.35** (0.19,0.57)	**0.33** (0.26,0.41)	**0.43** (0.35,0.51)	**0.23** (0.14,0.34)	**0.46** (0.34,0.61)
Hypoplastic Left Heart	**0.33** (0.17,0.57)	0 termin.	**0.23** (0.11,0.42)	**0.25** (0.13,0.45)	**0.23** (0.18,0.30)	**0.25** (0.20,0.32)	**0.20** (0.12,0.30)	**0.30** (0.21,0.43)
Cleft Palate	**1.03** (0.73,1.42)	0 termin.	**0.30** (0.16,0.51)	0 termin	**0.79** (0.69,0.91)	**0.86** (0.75,0.98)	**0.60** (0.46,0.77)	**0.69** (0.54,0.87)
Cleft Lip	**0.90** (0.62,1.26)	0 termin	**0.58** (0.37,0.85)	0 termin.	**0.70** (0.60,0.81)	**0.72** (0.62,0.83)	**0.59** (0.45,0.76)	**0.71** (0.55,0.89)
Esophageal Atresia	**0.22** (0.09,0.43)	0 termin	**0.21** (0.10,0.39)	0 termin.	**0.25** (0.19,0.32)	**0.26** (0.20,0.33)	**0.24** (0.15,0.35)	**0.25** (0.17,0.37)
Anal Atresia	**0.41** (0.23,0.67)	**0.52** (0.31,0.81)	**0.35** (0.19,0.57)	0 termin	**0.32** (0.26,0.40)	**0.35** (0.28,0.43)	**0.26** (0.17,0.37)	**0.28** (0.19,0.41)
Renal Agenesis	**0.38** (0.21,0.64)	**0.43** (0.25,0.70)	**0.30** (0.16,0.51)	**0.35** (0.19,0.57)	**0.18** (0.13,0.24)	**0.22** (0.16,0.28)	**0.01** (0.00,0.05)	**0.16** (0.09,0.25)
Limb Reductions	**0.63** (0.40,0.94)	**0.68** (0.44,1.00)	**0.49** (0.30,0.74)	**0.51** (0.32,0.77)	**0.42** (0.34,0.51)	**0.48** (0.40,0.57)	**0.40** (0.29,0.55)	**0.51** (0.38,0.67)
Abdominal Wall Defects	**0.73** (0.48,1.07)	**0.82** (0.55,1.16)	**0.28** (0.14,0.48)	**0.35** (0.19,0.57)	**0.39** (0.32,0.48)	**0.47** (0.39,0.56)	**0.30** (0.21,0.43)	**0.41** (0.30,0.56)
Down Syndrome	**1.11** (0.80,1.51)	**1.52** (1.15,1.97)	**1.06** (0.78,1.42)	**1.22** (0.92,1.60)	**1.09** (0.96,1.22)	**1.55** (1.40,1.71)	**0.88** (0.71,1.09)	**2.15** (1.87,2.45)

*International Clearinghouse of Birth Defects Monitoring Systems
L = livebirth S = stillbirth T = termination
‡ Total births = Live births + stillbirths

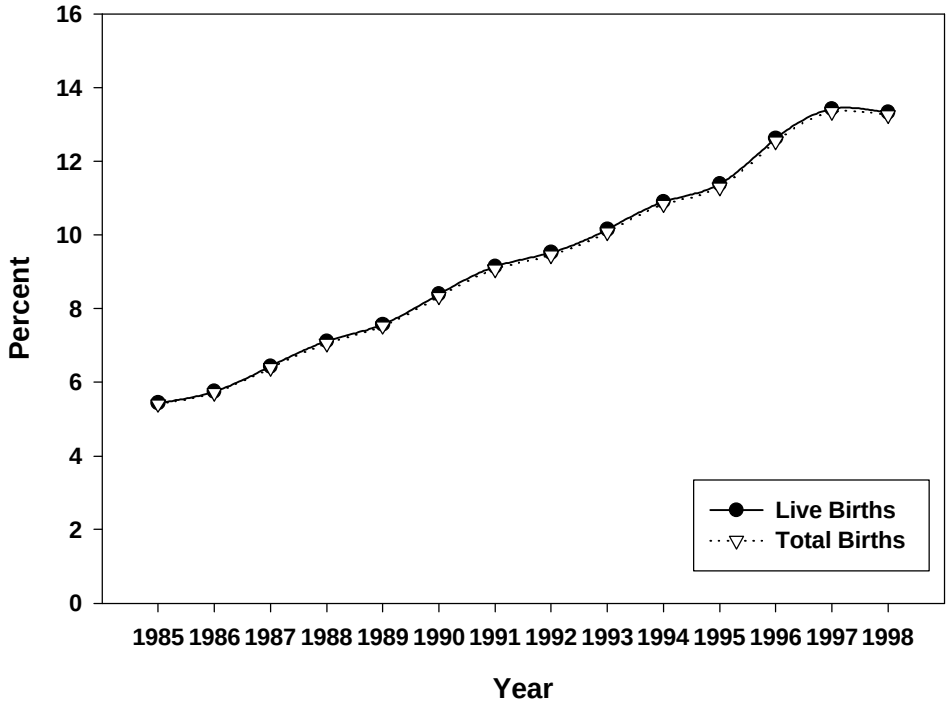
Table A7.2 Rates of selected congenital anomalies per 1,000 total ‡ births with 95 per cent confidence intervals.

Anomaly	England -Wales L,S	England -Wales L,S,T	Finland L,S	Finland L,S,T	Hungary L,S	Hungary L,S,T	Norway L,S	Norway L,S,T
Anencephaly	**0.04** (0.03,0.06)	**0.28** (0.24,0.33)	**0.08** (0.03,0.19)	**0.27** (0.15,0.43)	**0.04** (0.01,0.10)	**0.19** (0.11,0.29)	**0.12** (0.05,0.24)	**0.22** (0.12,0.37)
Spina Bifida	**0.07** (0.05,0.10)	**0.23** (0.20,0.27)	**0.40** (0.26,0.60)	**0.54** (0.37,0.76)	**0.13** (0.07,0.22)	**0.33** (0.23,0.46)	**0.27** (0.15,0.43)	**0.38** (0.24,0.57)
Encephalocele	**0.01** (0.00,0.02)	**0.04** (0.02,0.05)	**0.07** (0.02,0.17)	**0.18** (0.09,0.33)	**0.01** (0.00,0.05)	**0.02** (0.00,0.07)	**0.03** (0.00,0.11)	0 termin.
All NTDs	**0.13** (0.10,0.16)	**0.55** (0.50,0.61)	**0.55** (0.38,0.78)	**0.99** (0.75,1.27)	**0.18** (0.11,0.28)	**0.53** (0.40,0.70)	**0.42** (0.27,0.62)	**0.63** (0.45,0.87)
Hydrocephaly	**0.09** (0.07,0.12)	**0.18** (0.15,0.22)	**0.37** (0.23,0.56)	**0.50** (0.34,0.72)	**0.21** (0.13,0.32)	**0.38** (0.27,0.52)	**0.35** (0.22,0.54)	**0.37** (0.23,0.55)
Hypoplastic Left Heart	**0.04** (0.03,0.06)	**0.08** (0.06,0.10)	**0.24** (0.13,0.39)	0 termin.	**0.03** (0.01,0.08)	**0.07** (0.03,0.14)	**0.15** (0.07,0.28)	**0.18** (0.09,0.33)
Cleft Palate	**0.31** (0.26,0.35)	**0.31** (0.26,0.35)	**1.31** (1.04,1.63)	**1.32** (1.05,1.65)	**0.21** (0.13,0.32)	0 termin.	**0.72** (0.52,0.97)	0 termin.
Cleft Lip	**0.58** (0.52,0.64)	**0.58** (0.53,0.64)	**1.16** (0.90,1.47)	**1.25** (0.99,1.57)	**0.48** (0.35,0.63)	0 termin.	**1.33** (1.06,1.66)	**1.35** (1.07,1.68)
Esophageal Atresia	**0.07** (0.05,0.09)	0 termin.	**0.29** (0.17,0.46)	**0.32** (0.19,0.50)	**0.09** (0.04,0.17)	0 termin.	**0.10** (0.04,0.22)	0 termin.
Anal Atresia	**0.12** (0.10,0.15)	0 termin.	**0.39** (0.25,0.58)	**0.43** (0.28,0.64)	**0.09** (0.04,0.17)	0 termin.	**0.15** (0.07,0.28)	**0.18** (0.09,0.33)
Renal Agenesis	**0.05** (0.03,0.07)	**0.10** (0.08,0.13)	**0.12** (0.05,0.24)	**0.22** (0.12,0.37)	**0.02** (0.00,0.07)	0 termin.	**0.12** (0.05,0.24)	**0.15** (0.07,0.28)
Limb Reductions	**0.21** (0.17,0.24)	**0.23** (0.20,0.27)	**0.50** (0.34,0.72)	**0.65** (0.46,0.89)	**0.27** (0.18,0.39)	**0.29** (0.19,0.41)	**0.57** (0.39,0.79)	**0.60** (0.42,0.83)
Abdominal Wall Defects	**0.20** (0.17,0.24)	**0.23** (0.20,0.27)	**0.35** (0.22,0.54)	**0.55** (0.38,0.78)	**0.10** (0.05,0.18)	**0.19** (0.11,0.29)	**0.50** (0.34,0.71)	0 termin
Down Syndrome	**0.43** (0.38,0.49)	**0.93** (0.85,1.00)	**1.01** (0.77,1.30)	**2.29** (1.92,2.71)	**0.37** (0.26,0.51)	**0.54** (0.41,0.71)	**1.05** (0.81,1.35)	**1.17** (0.91,1.47)

*International Clearinghouse of Birth Defects Monitoring Systems
L = livebirth S = stillbirth T = termination
‡ Total births = Live births + stillbirths

APPENDIX 8. BIRTHS TO WOMEN AGE 35 AND OLDER.

Table A8. Per cent of live births and total births to women age 35 and older in Alberta, 1985 – 1998.



Total births = live births + stillbirths
Does not include terminations of pregnancy.

APPENDIX 9. BIRTHS IN ALBERTA, 1980-1998

The following Alberta Birth Statistics were obtained from *Vital Statistics Annual Reviews*, Alberta Government Services, Registries and are for Alberta residents only. The values under Section 1 were used as denominators for the rate and confidence interval calculations found in Tables A2, A3 and A4. The male birth numbers in Table A9.3 were used as denominators for the rate and confidence interval calculations for ICD-9/BPA codes 7525, 7526 and 7528 (Table A2). The female birth numbers in Table A9.2 were used as denominators for the rate and confidence interval calculations for ICD-9/BPA codes 7520, 7521, 7522, 7523 and 7524 (Table A2). Until 1991, Vital Statistics provided only total stillbirth numbers. Since 1991, the stillbirth numbers have been available by sex as well.

Table A9.1 Total Births in Alberta, 1980 – 1998.

	1980-1990	1991-1995	1996	1997	1998	1991-1998	1980-1998
FEMALE LB	228 978	98 540	18 074	17 801	18 284	152 699	381 677
MALE LB	241 326	103 376	19 400	18 749	19 333	160 851	402 184
STILL	3 211	1 380	235	248*	189	2 052	5 263
TOTAL	473 515	203 296	37 709	36 798	37 808	315 611	789 126

* 3 sex unknown

Table A9.2 Number of Female Births in Alberta, 1980 – 1998.

	1980-1990	1991-1995	1996	1997	1998	1991-1998	1980-1998
FEMALE LB	228 978	98 540	18 074	17 801	18 284	152 699	381 677
FEMALE SB	N/A	658	116	121*	90	985	985†
TOTAL	228 978‡	99 198	18 190	17 922	18 374	153 684	382 662†

* excluding sex unknown ‡ no stillbirths included in total † includes stillbirths from 1991-1998 only

Table A9.3 Total number of Male Births in Alberta, 1980 –1998.

	1980-1990	1991-1995	1996	1997	1998	1991-1998	1980-1998
MALE LB	241 326	103 376	19 400	18 749	19 333	160 858	402 184
MALE SB	N/A	702	119	124*	99	1 044	1 044†
TOTAL	241 326‡	104 078	19 519	18 873	19 432	161 902	403 228†

* excluding sex unknown ‡ no stillbirths included in total † includes stillbirths from 1991-1998 only

APPENDIX 10. LIVE BIRTHS AND STILLBIRTHS BY REGIONAL HEALTH AUTHORITY

Table A10 Number of live births and stillbirths per Regional Health Authority, 1996-1998.

Regional Health Authority	1996			1997			1998		
	Live	Still	Total	Live	Still	Total	Live	Still	Total
1. Chinook	2150	14	2164	2243	9	2252	2078	3	2081
2. Palliser	1161	8	1169	1183	9	1192	1242	7	1249
3. Headwaters	847	10	857	910	3	913	887	12	899
4. Calgary	11152	72	11224	11053	79	11132	11602	62	11664
5. Health Authority 5	650	0	650	622	4	626	625	6	631
6. David Thompson	2586	15	2601	2499	19	2518	2555	11	2566
7. East Central	1058	7	1065	1009	12	1021	1037	9	1046
8. WestView	1150	7	1157	1152	10	1162	1139	7	1146
9. Crossroads	985	2	987	934	5	939	564	1	565
10. Capital	9744	59	9803	9177	56	9233	9963	42	10005
11. Aspen	1191	6	1197	1098	8	1106	1125	4	1129
12. Lakeland	1503	13	1516	1496	8	1504	1523	6	1529
13. Mistahia	1322	10	1332	1273	10	1283	1327	3	1330
14. Peace	331	2	333	294	3	297	320	1	321
15. Keeweetinok	529	3	532	500	7	507	521	7	528
16. Northern Lights	599	3	602	568	5	573	590	3	593
17. Northwestern	518	5	523	539	2	541	508	6	514
Unknown	0	0	0	0	0	0	52	7	59
ALBERTA TOTAL	**37476**	**236**	**37712**	**36550**	**249**	**36799**	**37658**	**197**	**37855**

Source of Data: Alberta Vital Statistics Birth Registration and Stillbirth Registration

APPENDIX 11. REQUESTS TO ACASS FOR DATA

1997

- Medical Genetics Clinic, Calgary: Requested number of cases of Down Syndrome, and number of births to women over 35 years of age.
- International Clearinghouse of Birth Defects Monitoring Systems (ICBDMS): Requested 1995 sentinel defect data for their Annual Report.
- Medical Genetics Clinic, Calgary: Requested number of cases of Fetal Alcohol Syndrome from 1980-1995.
- Cardiologist, Alberta Children's Hospital: Requested number of cases of congenital heart defects in southern Alberta, 1990-1995.
- Poster presentation, Child health Research Symposium, Alberta Children's Hospital: Review of birth defect rates in Alberta, specifically anencephaly, spina bifida and Down Syndrome, 1980-1995.
- ICBDMS Annual meeting, Cape Town, South Africa: ACASS data on diaphragmatic hernia, sentinel defects for 1996 and number of stillbirths for which autopsies available.
- Headwaters Health Authority, Health Assessment Facilitator: Requested numbers and rates of cases with birth defects for 1994-1995 with brief outline of types of defects most commonly ascertained.
- Alberta Medical Association, Reproductive Care Committee: Requested 1995 sentinel defect rates and trends for inclusion in the Alberta Perinatal and Neonatal Statistics and Maternal Mortality Annual Report, 1995.

1998

- Medical Genetics Clinic, Calgary: Requested numbers and rates of anomalies of the diaphragm and diaphragmatic hernia from 1980-1997 in live births and stillbirths.
- ICBDMS: Requested 1996 sentinel defect data for their Annual Report.
- Post-RN student at the University of Calgary: Requested information about ACASS and data on major anomalies for class assignment.
- School Age Developmental Clinic, Alberta Children's Hospital: Requested data on the number of children born with Down Syndrome per year in Alberta.
- Medical Geneticist, Alberta Children's Hospital: Requested data on diaphragmatic hernia and anomalies of the diaphragm from 1980-1997.
- Paediatrician, Alberta Children's Hospital: Requested data on tracheo-esophageal fistula and esophageal atresia 1980-1995.
- Child Health Research Symposium, Alberta Children's Hospital: Poster and presentation.
- CDC, Atlanta: Request for data on hypospadias and cryptorchidism for 1980-1996.
- Mistahia Regional Health Authority: Requesting data on congenital anomalies for the region through 1995.
- Medical Genetics Clinic, Alberta Children's Hospital: Request for data on Glutaric Aciduria, 1990-1996.
- Calgary Between Friends, a recreational and social programme for youth with disabilities: Requested data on Down Syndrome for Calgary and Alberta for 1987-1996 inclusive.
- East Central Regional Health Authority: Request for data on congenital anomalies for 1987-1996 inclusive for health promotion planning.
- Physician, Department of Community Health Sciences, University of Calgary: Request for information and data for course preparation.

- Lethbridge Genetics Outreach: Requested data on seasonal variation of clefting.
- Paediatrician, Mistahia Regional Health Authority: Requested data on congenital heart defects in the region.
- Medical Geneticist, Department of Medical Genetics, Alberta Children's Hospital: Requested data on FAS/FAE.

1999

- Medical Geneticist, Department of Medical Genetics, Alberta Children's Hospital: Requested data on seasonal variation of a number of birth defects.
- ICBDMS: Requested 1997 sentinel defect data for their Annual Report.
- Cleft Palate Clinic, Alberta Children's Hospital: Requested information re bilateral cleft lip and palate rates.
- Pharmacy resident, Alberta Children's Hospital: Requested baseline data on birth defects and information about ACASS.
- Chinook Regional Health Authority: Requested data on congenital heart defects in the region through 1995.
- Medical Geneticist, Department of Medical Genetics, Alberta Children's Hospital: Requested data on Down Syndrome and omphalocele.
- Crossroads Regional Health Authority: Request for data on defects for 5 year strategic planning.
- Consultant working with Mistahia Regional Health Authority: Requested data for health indicators for follow-up of needs assessment.
- Investigation into congenital heart defects in Mistahia Region by Dr. RB Lowry, MD, medical Consultant for ACASS.
- Medical Geneticist, Department of Medical Genetics, Alberta Children's Hospital: Request for data on omphalocele and gastroschisis for 1993-1997 inclusive.
- Paediatric Cardiologist, Edmonton: Request for data on congenital heart defects – rates, proportion of all congenital anomalies, rates of all congenital anomalies in Alberta.
- Pulmonary Fellow, Alberta Children's Hospital: Requested data on tracheo-esophageal fistula and esophageal atresia.
- Medicine Hat Genetics Outreach: Requested data on Down Syndrome rates for the region and for the province.
- Obstetrician, Foothills Medical Centre/University of Calgary: Requested data on hypospadias and epispadias for 1980-1997 inclusive.
- Fetal Centre, Hospital for Sick Children, Toronto: Requested information on ACASS and how system operates – e.g. ascertainment, coding, forms.

2000

- Medical Geneticist, Department of Medical Genetics, Alberta Children's Hospital: Requested data on congenital foot anomalies.
- ICBDMS: Requested 1998 sentinel defect data for their Annual Report.
- Medical Geneticist, Department of Medical Genetics, Alberta Children's Hospital: Requested data on anomalies among Hutterites in Alberta.
- Medical Geneticist, Department of Medical Genetics, Alberta Children's Hospital: Requested data on Pierre Robin Sequence.
- University student: Requested data on Cri du Chat Syndrome.
- Obstetrician, Foothills Medical Centre/University of Calgary: Requested data on sex ratio of fetal cases and cytogenetic abnormalities.
- CNIB: Requested data on ocular albinism in southern Alberta.

- University student: Enquired about the Surveillance system.
- Parent of a child with congenital anomaly: Requested rates of limb reductions in Alberta.
- Antenatal Genetics Clinic, Department of Medical Genetics, Alberta Children's Hospital: Requested data on omphalocele and NTDs.
- Medical Geneticist, Department of Medical Genetics, B.C. Children's Hospital: Requested data on abdominal wall defects and hernias.
- Medical Geneticist, Department of Medical Genetics, University of Alberta Hospitals: Requested data on Hutterite births.
- Medical Geneticist, Department of Medical Genetics, Alberta Children's Hospital: Requested data on Hutterites and cystic fibrosis.
- Alberta Health and Wellness: Requested data on Prader-Willi Syndrome in response to newspaper article on the topic.
- Medical Geneticist, Department of Medical Genetics, Alberta Children's Hospital: Requested data on gastroschisis and maternal age.
- Antenatal Genetics Clinic, Department of Medical Genetics, Alberta Children's Hospital: Requested data on Down Syndrome and maternal age.
- Genetic Counsellor, Department of Medical Genetics, Alberta Children's Hospital: Requested data on cleft lip and palate and neural tube defects.
- Lethbridge Cerebral Palsy Association: Requested data on cerebral palsy in Alberta.
- Presentation to Canadian Congenital Anomalies Surveillance System: Re ACASS and trends in Alberta.
- Presentation to Medical Genetics Residents: Re ACASS e.g. ascertainment, trends.