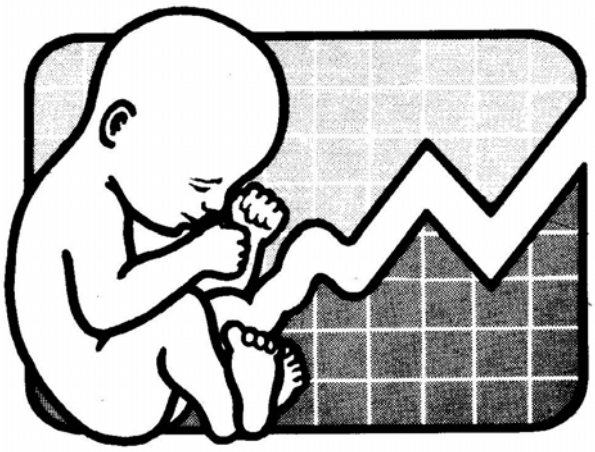
Arizona Department of Health Services

1998 - 2000
Arizona Birth Defects Monitoring Program Report



Janet Napolitano
Governor
State of Arizona

Susan Gerard
Director
Arizona Department of Health Services

Dear Residents of Arizona,

We are please to present you with the 'Arizona Birth Defects Monitoring Program Report' covering the years from 1998 to 2000. The results from this monitoring program illuminate opportunities for increased prevention and treatment of birth defects, which will enable us to work towards our vision of providing leadership for a healthier Arizona.

We would like to thank all the individuals who contributed to and produced this informative report. We sincerely hope that the information in this report helps you, our partners, in your work to improve the health of all Arizonans.

1998 - 2000
ARIZONA BIRTH DEFECTS MONITORING PROGRAM REPORT

Arizona Birth Defects Monitoring Program
Office of Health Registries
Bureau of Public Health Statistics
Arizona Department of Health Services

150 North 18th Avenue, Suite 550
Phoenix, Arizona 85007
(602) 542-7308

November 30, 2006

The Arizona Department of Health Services does not discriminate on the basis of disability in the administration of its programs and services as prescribed by Title II of the Americans with Disabilities Act of 1990 and Section 504 of the Rehabilitation Act of 1973.

If you need this publication in an alternative format, please contact the ADHS Office of Health Registries, Bureau of Public Health Statistics at (602) 542-7308.

Suggested Citation: *1998 to 2000 Arizona Birth Defects Monitoring Program Report*. Arizona Department of Health Services, Phoenix, 2006. (Produced as a cooperative effort within the Arizona Birth Defects Monitoring Program.)

Staff

Richard S. Porter ..... Chief, Bureau of Public Health Statistics

Timothy Flood, M.D. ..... Medical Director, Bureau of Public Health Statistics

Georgia Armenta Yee, B.S.W., C.T.R ..... Chief, Office of Health Registries

Allison Varga James, R.N ..... Health Program Manager, ABDMP

BichTram Nguyen, B.S ..... Epidemiology Specialist, ABDMP

Hoa Tran, M.P.H. ..... Seasonal Health Program Manager, ABDMP

Matilda Armenta ..... Birth Defect Surveillance Team Leader

Amie Bobo ..... Birth Defect Data Specialist

Christina Gutierrez ..... Birth Defect Data Specialist

We thank the hospitals and health care providers of the state for their cooperation and active support of the Arizona Birth Defects Monitoring Program (ABDMP). Our special thanks go to the directors and employees of the medical record departments of the hospitals and clinics, without whose help we could not conduct our case-finding and abstraction.

TABLE OF CONTENTS

Tables

TABLE OF CONTENTS

Figures

Appendices

EXECUTIVE SUMMARY

The Arizona Birth Defects Monitoring Program (ABDMP) is a population-based registry which provides information on the occurrence of birth defects throughout the state of Arizona. This report includes information about the occurrence of 44 categories of structural birth defects diagnosed in children born to Arizona residents from 1998-2000 (see Appendix A for the definitions of birth defects reported by the ABDMP). The specific birth defects included in this report are significant because they require medical or surgical intervention, considerably affect the child's appearance, and/or seriously affect the health and development of the child.[1]

General Conclusions

- There were 4,516 infants born with reportable birth defects from 1998-2000 (approximately 1,500 infants/year).
- Two out of every 100 infants born in Arizona were diagnosed with a reportable birth defect before their first birthday.
- There were 1,886 stillbirths from 1998-2000; nine out of every 100 stillborns had a reportable birth defect.
- The incidence rates of reportable birth defects were similar to what they had been in previous years.
- The most common birth defects were hypospadias and epispadias, pyloric stenosis, heart defects, obstructive genitourinary defects, Down syndrome, and cleft lip with or without cleft palate.

Findings of Significance

Neural Tube Defects

- There were 180 infants born with neural tube defects (NTDs) in this three year period. Neural tube defects often cause paralysis, severe mental retardation, or death. Evidence shows that 50% to 70% of these kinds of birth defects can be prevented if all women of childbearing age take 400 mcg of folic acid daily, starting before they become pregnant.[2,3,4]
- There was a 26% decrease in NTD-affected pregnancies in the United States from 1995-1996 to 1999-2000. There was a 16% decline in NTD rates during the same time period in Arizona. This decrease is associated with the Food and Drug Administration's mandate that all cereal grains be fortified with folic acid starting in 1998.[5]
- Many more NTDs may be prevented if the following actions are taken:
  - Develop and implement strategies to increase the percentage of women of childbearing age consuming 400 mcg of folic acid daily.
  - Encourage primary health care providers to discuss the importance of folic acid intake with all women of childbearing age.
  - Encourage obstetricians to ensure that women scheduling their first appointment for prenatal care are instructed to start or continue taking a

prenatal vitamin containing folic acid daily.

- Encourage all corn flour producers to fortify their products with folic acid.
- Encourage the federal government to increase the level of folic acid fortification in wheat products.
- Continue programs that provide low income women of childbearing age and their spouses with folic acid education and that distribute multivitamins containing folic acid to women.
- Ensure all women receive adequate preconceptional and prenatal healthcare to provide education on behaviors to minimize the risk of birth defects.

Gastroschisis and Heart Defects

- The rate of gastroschisis among Native Americans (7 per 10,000 live births) was significantly higher than the rates for Whites (4 per 10,000 live births) and Hispanics (5 per 10,000 live births).
- The rate of gastroschisis for births to mothers less than 20 years of age (14 per 10,000 live births) was significantly higher than the state rate (5 per 10,000 live births) for gastroschisis.
- The heart defect rates for Gila County (103 per 10,000 live births), Navajo County (76 per 10,000 live births) and Pima County (69 per 10,000 live births) were significantly higher than the state rate for heart defects (65 per 10,000 live births).
- Because of the high incidence rate of these defects in certain rural counties, it is important that rural health care systems are equipped to provide appropriate medical support (special education for providers, up to date technology, transport services to higher level facilities, and consultation resources) for these medically fragile children in an effort to minimize the consequences of such defects.

Birth defects affect thousands of families in the state of Arizona. The Arizona Birth Defects Monitoring Program plays a vital role in accurately identifying birth defects that occur throughout the state and provides this data to state and community leaders and health care professionals so that they can plan, implement, and evaluate programs for prevention and treatment of birth defects in Arizona.

THE IMPORTANCE OF ARIZONA'S BIRTH DEFECTS REGISTRY

Introduction

Birth defect surveillance programs in the United States were first established in response to concerns over environmental pollutants and the finding that severe birth defects were associated with pregnant women who, without knowing the harmful effects on the fetus, used medications such as thalidomide. At present, birth defect surveillance programs are being used to monitor trends in birth defect rates over time, to evaluate birth defect prevention programs, and facilitate research efforts to identify the etiology of birth defects.[6,7]

The Arizona Birth Defects Monitoring Program (ABDMP) is a population-based registry which provides information on the occurrence of 44 categories of birth defects. These specific birth defects are monitored at the recommendation of the National Birth Defects Prevention Network and Centers for Disease Control and Prevention. They were selected because of a combination of factors: they require medical or surgical intervention, they considerably affect the child's appearance, they seriously affect the health and development of the child, they have a significant public health impact, their frequency of occurrence, and/or the level of existing knowledge on their etiology and risk factors.[8] The ABDMP provides ongoing surveillance to monitor trends in the occurrence of these birth defects and detect the onset of possible problems.[9,10,11] The information is used to plan and evaluate birth defect prevention efforts and to direct allocation of resources for health services. Such a registry is necessary because other systems for reporting birth defects, such as birth certificates and hospital discharge data, tend to be inaccurate or incomplete due to under-reporting of cases, lack of specificity of the type of birth defect, and/or incomplete demographic data.[12]

To provide accurate and current information for health planning and prevention activities, in 2003 the ABDMP entered into a five-year Cooperative Agreement with the Centers for Disease Control and Prevention to develop a rapid reporting system (RRS) for birth defects. The goal of the RRS is to identify infants born in Arizona with anencephaly, encephalocele, spina bifida, cleft lip, or cleft palate within six months of birth and to provide families of living children with information and resources for follow-up medical and social services. In this way the ABDMP

hopes to increase the percentage of children with these birth defects who receive coordinated, multi-disciplinary follow-up services to minimize the long-term disabilities associated with their condition. It is hoped that this program will also increase the number of families of children with birth defects receiving support from other families of children with similar difficulties. Furthermore, this rapid-surveillance system will enable the Arizona Department of Health Services Folate Committee to track trends in neural tube defect (NTD) occurrence and evaluate the efficacy of programs educating the public on folic acid for NTD prevention.

Economic Cost

Birth defects are expensive, both in their costs in human productivity and monetary resources. They remain the leading cause of infant mortality in both Arizona and the United States. Arizona and national data show that between 1989 and 2003, 20-25% of infant deaths were due to a birth defect. In comparison, 13-17% of infant deaths in 2003 were attributed to low birth weight and 5-8% were attributed to SIDS.[13,14] Birth defects are also the fifth leading cause of years of potential life lost.[15] Although many infants with birth defects survive beyond their first birthday, many require special medical services, education and rehabilitation services, vocational training and/or custodial care. Lifetime costs for these services is estimated to be $75,000 to over $500,000 per child.[16] These costs contribute to a lifetime of hardship for these children, their families, and their communities.

Known Causes of Birth Defects

Genetic and environmental factors can cause birth defects. There are three major categories of known causes of birth defects. The first category is "chromosomal errors," such as Down syndrome. The second is "environmental factors," such as maternal alcohol consumption, which may cause fetal alcohol syndrome. The third category is "maternal illness, infections, or medical conditions," such as German measles, which may cause congenital rubella. In spite of research efforts in identifying the etiology of birth defects, the causes of most birth defects remain unknown.. This is suspected to be because most birth defects are caused by complex combinations of genetic and environmental factors that are very difficult to identify – particularly in studies of relatively rare conditions.[17, 18, 19]

Interventions

Although the causes of birth defects are largely unknown, there are many important steps women can take to minimize the risk of such defects. For example, in recent years, maternal intake of the B-vitamin folic acid has been shown to be necessary for proper fetal development. Several prospective case-controlled studies have shown that the consumption of 400 mcg of folic acid daily starting prior to conception and through the first trimester has reduced the incidence of neural tube defects (NTDs) by at least 50%.[20] There is also research that shows that folic acid may reduce the incidence of certain types of heart defects, urinary tract defects, and oral clefts.[21,22,23] A considerable amount of research still needs to be undertaken to augment our knowledge of birth defects, their etiology, and their impact on different subpopulation groups. If Arizona is to ensure the well-being of its children, it is essential that the surveillance and documentation of the occurrence of birth defects in the state be undertaken and made available to the public and researchers.

METHODS

The ABDMP is a statewide, population-based, active ascertainment program, pursuant to Arizona Revised Statute §36-133, which mandates the surveillance of chronic diseases, including birth defects. The funding for the ABDMP comes from appropriations of the Arizona state legislature, monies collected through the Arizona Newborn Screening Program, and federal funds from the Maternal Child Health Block Grant and a Cooperative Agreement with the Centers for Disease Control and Prevention. Trained ABDMP staff members collect data from 65 reporting sources including: 58 hospitals, two centers providing genetics services, and four state Children's Rehabilitative Services clinics. Ascertainment procedures used by the ABDMP are nearly identical to those used by the U.S. Centers for Disease Control's Metropolitan Atlanta Congenital Defects Program, the gold standard for birth defect registries internationally.[24]

Hospital case-finding sources include disease indexes, labor and delivery logs, nursery logs, newborn intensive care unit logs, pediatric logs, and autopsy logs. Potential cases are also identified through a review of the Hospital Discharge Data Base and birth and fetal death certificates. All records for children up to one year of age with recorded birth defects become part of this listing of possible cases. The medical records of possible cases are reviewed to determine which records meet the case definition. An abstract (case report) of the medical record is completed for each reportable case. If the nature of a defect diagnosed in the first year of life is more precisely diagnosed later in the child's life and this information is contained in the chart at the time of our review, then the more precise diagnosis is used. ABDMP staff then assign a six-digit classification code to each reportable defect. The classification system is CDC's modification of the British Pediatric Association (BPA) Classification of Disease. BPA coding is an extension of the World Health Organization's International Classification of Disease, 9th Revision, Clinical Modification (ICD-9-CM). (See Appendix B for a list of BPA codes used by the ABDMP.) The abstracts of cases identified from multiple sources are compared, merged, and added to the registry. To identify children born with birth defects in the year 2000, the ABDMP staff reviewed 6,092 medical records. 2,249 of these medical records contained information concerning 1,600 infants that met the case definition (many of whom were identified in medical records at more than one hospital or clinic).

CASE DEFINITION

The following are the criteria for inclusion in the Birth Defects Monitoring Program case file:

A. The mother's place of residence at the time of birth must be in Arizona.

B. The child must have a birth defect that is reported by the ABDMP (see Appendix B for the list of reported birth defects).

C. The health care provider must have had a high level of confidence in the diagnosis (defined as a precision of diagnosis code of eight or greater, as explained in Appendix C.)

D. The defect must be diagnosed, or signs and symptoms of a potential defect recognized, within the first year of life.

E. The child must have an Arizona-issued birth or fetal death certificate.

F. The date of birth (or delivery for stillbirths > 19 weeks of gestational age) must be on or after January 1, 1986.

F. For a list of exclusions, see Appendix D.

Due to the need to collect and report data on birth defects in a more timely manner, effective March 1996, the ABDMP reduced the number of reportable conditions from over 500 to 140. The 140 conditions, divided into 44 categories of defects, include only the major congenital anomalies recommended by The International Clearinghouse for Birth Defects Monitoring Systems and the Centers for Disease Control and Prevention (CDC). The 140 conditions still permit the ABDMP to compare its rates with other registries for the major birth defect categories. The reduced list of reportable defects applies to data collected on births after 1991.

For the first time, data for the ABDMP Report (for 1986-2000 cases) has been analyzed using birth defect categories as defined by the CDC. (See Appendix B.) Data for previous reports were analyzed using birth defect categories defined by the ABDMP. This change only affects the reporting of anencephaly and spina bifida cases.

INTERPRETING THE DATA

The tables and figures presented in this report represent data collected on birth defects in Arizona for the period 1986 to 2000. Each table presents the reported counts, rates, and confidence intervals on selected congenital anomalies. Below is an explanation of how counts, rates, and confidence intervals were calculated.

Counts

The counts, sometimes called cases, represent the number of children who, in the first year of life, were diagnosed with at least one reportable birth defect within the defect category. Children born with more than one reportable defect, as often occurs, may be counted in more than one category.

Rates

Incidence rates of birth defects were calculated by dividing the number of children (cases) with a particular reportable defect by the total number of live births (and in some cases live births plus fetal deaths) for the specific year of interest, and then multiplying by 10,000. Most tables and figures show rates that are calculated by including live births and fetal deaths in the numerator and only live births for the denominator (the inclusion of fetal deaths in the denominator does not change the rates significantly). For example:

$$\frac{110 \textit{ live borns or stillborns with Down syndrome born in the year } 2000}{84{,}866 \textit{ live births in the year } 2000} * 10{,}000 = \quad \textit{12.96 cases of Down syndrome per 10,000 live births}$$

Confidence Intervals

The confidence intervals shown in the tables and figures are provided to give information about the estimate of the rate. Confidence intervals presented in this report are 95 percent Poisson confidence intervals. The confidence intervals indicate that the true rate should be contained in this interval 95 percent of the time. For example, Down syndrome in the year 2000 occurs at a rate of 12.96 per 10,000 births. The lower and upper bounds of the point estimate of this rate are 10.5 and 15.4, respectively. Thus, one can say that 95 percent of the time the true rate of Down syndrome is between 10.5 and 15.4 cases per 10,000 live births and fetal deaths. The following

formula was use to calculate the confidence intervals. Confidence Interval = Rate ± 1.96√Var (Var = (Rate / Population) x 10,000).

Small Numbers and a Note Of Caution

While the intent of these data is to provide useful information on birth defects in Arizona, it is equally important not to mislead data users. Rates, confidence intervals, and any other analyses based on fewer than 10 reported cases cannot be considered statistically reliable and are not shown for local areas.

Tests of Significance

Z tests were used to determine whether there were statistically significant differences in the rates between groups and areas. The state rate was used as the standard rate in these tests.

STATE PROFILE OF DEFECTS

The Arizona Birth Defects Monitoring Program (ABDMP) has been in operation since 1986. This is the twelfth report of data compiled by the ABDMP in its mission to collect, analyze, and disseminate information on children with birth defects.

Tables and Figures

Table 1-A (pages 11-12) presents data on the 44 categories of birth defects collected by the ABDMP among live born and stillborn infants born statewide, analyzed by race/ethnicity, for 1998. Tables 1-B (pages 13-14) and 1-C (pages 15-16) present similar data for 1999 and 2000. Tables 2-A (page 17), 2-B (page 18), and 2-C (page 19) display the number of live born and stillborn infants with all reportable birth defects and the average number of defects among live born and stillborn infants analyzed by county of maternal residence for 1998, 1999, and 2000, respectively. Table 3 (pages 20-22) displays the number of cases and the rates of the 44 categories of anomalies by year for 1986 through 2000. The series of graphs in Figure 1 (pages 23-30) display the trends for 1986 through 2000 for the same categories of defects.

Neural Tube Defects

Figure 2 (page 31) shows neural tube defects rates (combining data for anencephaly, encephalocele, and spina bifida) for 1986-2000.

Race/Ethnicity

Spina bifida, gastroschisis, and omphalocele rates have been stratified by race and ethnicity in Figure 3 (page 33) and Figure 4 (page 34).

Maternal Age

Figure 5 (page 35) compares rates of gastroschisis occurring in 1997-2000 among different maternal age groups.

County Profiles

Tables 4-7 (pages 37-40) present aggregated data on the number of infants with neural tube defects, gastroschisis, omphalocele, and heart defects born between 1986 to 2000 in each county.

Table 1 - A

Arizona Birth Defects Monitoring Program

Live-born and Stillborn Cases of Congenital Anomalies by Race/Ethnicity - Arizona 1998

Incidence Rate [a,b] Per 10,000 Live Births

CONDITION	TOTAL[c]	RATE	WHITE NON-HISP.	RATE	HISP.	RATE	BLACK	RATE	NATIVE AMER.	RATE	OTHER	RATE
Infants with at least one reportable defect[d]	1390	178.33	711	187.52	485	167.56	35	135.03	115	205.39	44	149.15
Amniotic bands	14	1.80	6	1.59	7	2.42	0	0.00	1	1.79	0	0.00
Anencephaly	19	2.44	9	2.38	9	3.12	0	0.00	1	1.79	0	0.00
Aniridia	3	0.38	0	0.00	2	0.69	0	0.00	1	1.79	0	0.00
Anophthalmia/microphthalmia	13	1.67	3	0.79	8	2.77	0	0.00	2	3.58	0	0.00
Anotia/microtia	22	2.82	7	1.85	11	3.81	0	0.00	4	7.16	0	0.00
Aortic valve stenosis	26	3.34	13	3.44	6	2.08	1	3.88	5	8.94	1	3.25
Atrial septal defect	122	15.65	49	12.96	49	16.96	3	11.63	16	28.62	5	16.23
Biliary atresia	5	0.64	2	0.53	3	1.04	0	0.00	0	0.00	0	0.00
Bladder exstrophy	2	0.26	1	0.26	0	0.00	1	3.88	0	0.00	0	0.00
Choanal atresia	12	1.54	6	1.59	3	1.04	0	0.00	1	1.79	2	6.49
Cleft lip with or without cleft palate	95	12.19	42	11.11	37	12.81	3	11.63	8	14.31	5	16.23
Cleft palate without cleft lip	53	6.80	24	6.35	16	5.54	2	7.75	9	16.10	2	6.49
Coarctation of aorta	37	4.75	16	4.23	14	4.85	0	0.00	6	10.73	1	3.25
Common truncus	11	1.41	5	1.32	4	1.38	0	0.00	2	3.58	0	0.00
Congenital cataract	11	1.41	3	0.79	3	1.04	0	0.00	3	5.37	1	3.25
Congenital hip dislocation	63	8.08	30	7.94	21	7.27	1	3.88	9	16.10	2	6.49
Diaphragmatic hernia	30	3.85	14	3.70	10	3.46	2	7.75	4	7.16	0	0.00
Down syndrome (Trisomy 21)	125	16.04	54	14.28	48	16.61	2	7.75	15	26.83	5	16.23
Ebstein's anomaly	5	0.64	1	0.26	4	1.38	0	0.00	0	0.00	0	0.00
Edwards syndrome (Trisomy 18)	12	1.54	6	1.59	5	1.73	0	0.00	1	1.79	0	0.00
Encephalocele	8	1.03	4	1.06	4	1.38	0	0.00	0	0.00	0	0.00
Endocardial cushion defect	44	5.65	21	5.56	19	6.58	1	3.88	2	3.58	1	3.25

[a] Incidence rates include live born and stillborn cases in numerator and only live borns in denominator.

[b] Incidence rates based on counts of less than 10 events are not statistically reliable.

[c] Total includes cases of unknown race/ethnicity, therefore it may be greater than the sum of the cases listed in the individual race/ethnicity categories

[d] Many infants have multiple defects and are included in multiple condition categories. Therefore, the numbers in this row are NOT column totals.

Table 1 – A (continued)

Arizona Birth Defects Monitoring Program

Live-born and Stillborn Cases of Congenital Anomalies by Race/Ethnicity - Arizona 1998

Incidence Rate [a,b] Per 10,000 Live Births

CONDITION	TOTAL[c]	RATE	WHITE NON-HISP.	RATE	HISP.	RATE	BLACK	RATE	NATIVE AMER.	RATE	OTHER	RATE
Esophageal atresia/Tracheoesophageal fistula	19	2.44	10	2.65	8	2.77	0	0.00	1	1.79	0	0.00
Fetus or newborn affected by maternal alcohol use	6	0.77	1	0.26	0	0.00	0	0.00	1	1.79	3	9.74
Gastroschisis	39	5.00	15	3.97	16	5.54	0	0.00	5	8.94	2	6.49
Hirschsprung disease	8	1.03	3	0.79	3	1.04	1	3.88	0	0.00	1	3.25
Hydrocephalus without spina bifida	44	5.65	21	5.56	13	4.50	2	7.75	4	7.16	4	12.98
Hypoplastic left heart syndrome	8	1.03	5	1.32	2	0.69	0	0.00	1	1.79	0	0.00
Hypospadias and epispadias	228	29.25	154	40.74	50	17.31	11	42.64	5	8.94	7	22.72
Microcephaly	53	6.80	18	4.76	21	7.27	1	3.88	7	12.52	5	16.23
Obstructive genitourinary defect	125	16.04	63	16.67	40	13.85	5	19.38	11	19.68	6	19.47
Omphalocele	14	1.80	8	2.12	5	1.73	0	0.00	0	0.00	1	3.25
Patau syndrome (Trisomy 13)	10	1.28	3	0.79	5	1.73	1	3.88	1	1.79	0	0.00
Pulmonary valve atresia and stenosis	56	7.18	31	8.20	23	7.96	0	0.00	2	3.58	0	0.00
Pyloric stenosis	135	17.32	83	21.96	48	16.61	0	0.00	3	5.37	1	3.25
Rectal and large intestinal atresia/stenosis	43	5.52	24	6.35	12	4.15	0	0.00	4	7.16	3	9.74
Reduction deformity, lower limbs	20	2.57	7	1.85	10	3.46	1	3.88	2	3.58	0	0.00
Reduction deformity, upper limbs	23	2.95	10	2.65	9	3.12	0	0.00	4	7.16	0	0.00
Renal agenesis/hypoplasia	36	4.62	23	6.08	8	2.77	1	3.88	3	5.37	0	0.00
Spina bifida without anencephalus	33	4.23	15	3.97	15	5.19	0	0.00	1	1.79	2	6.49
Tetralogy of Fallot	33	4.23	13	3.44	18	6.23	0	0.00	2	3.58	0	0.00
Transposition of great arteries	36	4.62	15	3.97	20	6.92	0	0.00	0	0.00	1	3.25
Tricuspid valve atresia and stenosis	7	0.90	4	1.06	2	0.69	0	0.00	1	1.79	0	0.00
Ventricular septal defect	127	16.29	45	11.90	60	20.77	4	15.50	14	25.04	4	12.98

[a] Incidence rates include live born and stillborn cases in numerator and only live borns in denominator.

[b] Incidence rates based on counts of less than 10 events are not statistically reliable.

[c] Total includes cases of unknown race/ethnicity, therefore it may be greater than the sum of the cases listed in the individual race/ethnicity categories

Table 1 - B

Arizona Birth Defects Monitoring Program

Live-born and Stillborn Cases of Congenital Anomalies by Race/Ethnicity - Arizona 1999

Incidence Rate [a,b] Per 10,000 Live Births

CONDITION	TOTAL[c]	RATE	WHITE NON-HISP.	RATE	HISP.	RATE	BLACK	RATE	NATIVE AMER.	RATE	OTHER	RATE
Infants with at least one reportable defect[d]	1427	176.37	681	178.59	527	170.48	38	143.02	114	203.10	67	186.42
Amniotic bands	10	1.24	6	1.57	3	0.97	1	3.76	0	0	0	0
Anencephaly	15	1.85	3	0.79	9	2.91	1	3.76	1	1.78	1	2.78
Aniridia	0	0	0	0	0	0	0	0	0	0	0	0
Anophthalmia/microphthalmia	14	1.73	7	1.84	6	1.94	0	0	1	1.78	0	0
Anotia/microtia	28	3.46	6	1.57	13	4.21	0	0	8	14.25	1	2.78
Aortic valve stenosis	42	5.19	25	6.56	13	4.21	0	0	3	5.34	1	2.78
Atrial septal defect	106	13.1	44	11.54	40	12.94	0	0	14	24.94	8	22.26
Biliary atresia	4	0.49	3	0.79	1	0.32	0	0	0	0	0	0
Bladder exstrophy	2	0.25	0	0	1	0.32	0	0	1	1.78	0	0
Choanal atresia	14	1.73	4	1.05	9	2.91	0	0	1	1.78	0	0
Cleft lip with or without cleft palate	90	11.12	44	11.54	31	10.03	2	7.53	8	14.25	5	13.91
Cleft palate without cleft lip	58	7.17	24	6.29	25	8.09	1	3.76	3	5.34	5	13.91
Coarctation of aorta	41	5.07	15	3.93	21	6.79	1	3.76	3	5.34	1	2.78
Common truncus	1	0.12	0	0	1	0.32	0	0	0	0	0	0
Congenital cataract	14	1.73	7	1.84	6	1.94	0	0	1	1.78	0	0
Congenital hip dislocation	57	7.05	23	6.03	29	9.38	0	0	5	8.91	0	0
Diaphragmatic hernia	19	2.35	9	2.36	7	2.26	1	3.76	1	1.78	1	2.78
Down syndrome (Trisomy 21)	115	14.21	48	12.59	47	15.2	2	7.53	12	21.38	6	16.69
Ebstein's anomaly	8	0.99	4	1.05	2	0.65	0	0	1	1.78	1	2.78
Edwards syndrome (Trisomy 18)	18	2.22	9	2.36	6	1.94	0	0	1	1.78	2	5.56
Encephalocele	11	1.36	6	1.57	3	0.97	1	3.76	1	1.78	0	0
Endocardial cushion defect	37	4.57	23	6.03	11	3.56	1	3.76	2	3.56	0	0

[a] Incidence rates include live born and stillborn cases in numerator and only live borns in denominator.

[b] Incidence rates based on counts of less than 10 events are not statistically reliable.

[c] Total includes cases of unknown race/ethnicity, therefore it may be greater than the sum of the cases listed in the individual race/ethnicity categories

[d] Many infants have multiple defects and are included in multiple condition categories. Therefore, the numbers in this row are NOT column totals.

Table 1 – B (continued)

Arizona Birth Defects Monitoring Program

Live-born and Stillborn Cases of Congenital Anomalies by Race/Ethnicity - Arizona 1999

Incidence Rate [a,b] Per 10,000 Live Births

CONDITION	TOTAL[c]	RATE	WHITE NON-HISP.	RATE	HISP.	RATE	BLACK	RATE	NATIVE AMER.	RATE	OTHER	RATE
Esophageal atresia / Tracheoesophageal fistula	14	1.73	5	1.31	4	1.29	2	7.53	2	3.56	0	0
Fetus or newborn affected by maternal alcohol use	12	1.48	2	0.52	1	0.32	0	0	6	10.69	2	5.56
Gastroschisis	28	3.46	10	2.62	13	4.21	0	0	3	5.34	2	5.56
Hirschsprung disease	13	1.61	6	1.57	3	0.97	1	3.76	0	0	3	8.35
Hydrocephalus without spina bifida	44	5.44	14	3.67	26	8.41	0	0	2	3.56	1	2.78
Hypoplastic left heart syndrome	13	1.61	6	1.57	5	1.62	0	0	1	1.78	1	2.78
Hypospadias and epispadias	234	28.92	142	37.24	63	20.38	11	41.4	11	19.6	6	16.69
Microcephaly	70	8.65	31	8.13	23	7.44	0	0	5	8.91	9	25.04
Obstructive genitourinary defect	122	15.08	52	13.64	51	16.5	1	3.76	15	26.72	3	8.35
Omphalocele	14	1.73	4	1.05	9	2.91	0	0	1	1.78	0	0
Patau syndrome (Trisomy 13)	8	0.99	3	0.79	3	0.97	1	3.76	0	0	1	2.78
Pulmonary valve atresia and stenosis	69	8.53	38	9.97	24	7.76	0	0	1	1.78	5	13.91
Pyloric stenosis	145	17.92	72	18.88	51	16.5	6	22.58	7	12.47	7	19.48
Rectal and large intestinal atresia/stenosis	28	3.46	14	3.67	11	3.56	1	3.76	2	3.56	0	0
Reduction deformity, lower limbs	17	2.1	5	1.31	5	1.62	1	3.76	4	7.13	2	5.56
Reduction deformity, upper limbs	29	3.58	17	4.46	10	3.23	0	0	1	1.78	1	2.78
Renal agenesis/hypoplasia	30	3.71	13	3.41	13	4.21	0	0	3	5.34	1	2.78
Spina bifida without anencephalus	40	4.94	19	4.98	16	5.18	1	3.76	2	3.56	1	2.78
Tetralogy of Fallot	25	3.09	14	3.67	6	1.94	1	3.76	1	1.78	3	8.35
Transposition of great arteries	39	4.82	23	6.03	12	3.88	2	7.53	0	0	2	5.56
Tricuspid valve atresia and stenosis	6	0.74	4	1.05	1	0.32	0	0	1	1.78	0	0
Ventricular septal defect	143	17.67	56	14.69	59	19.09	5	18.82	14	24.94	8	22.26

[a] Incidence rates include live born and stillborn cases in numerator and only live borns in denominator.

[b] Incidence rates based on counts of less than 10 events are not statistically reliable.

[c] Total includes cases of unknown race/ethnicity, therefore it may be greater than the sum of the cases listed in the individual race/ethnicity categories

Table 1 - C

Arizona Birth Defects Monitoring Program

Live-born and Stillborn Cases of Congenital Anomalies by Race/Ethnicity - Arizona 2000

Incidence Rate [a,b] Per 10,000 Live Births

CONDITION	TOTAL[c]	RATE	WHITE NON-HISP.	RATE	HISP.	RATE	BLACK	RATE	NATIVE AMER.	RATE	OTHER	RATE
Infants with at least one reportable defect[d]	1541	181.58	723	185.36	611	179.38	49	180.68	97	170.09	61	180.21
Amniotic bands	14	1.65	5	1.28	8	2.35	1	3.69	0	0	0	0
Anencephaly	14	1.65	5	1.28	5	1.47	2	7.37	0	0	2	5.91
Aniridia	1	0.12	1	0.26	0	0	0	0	0	0	0	0
Anophthalmia/microphthalmia	22	2.59	7	1.79	10	2.94	0	0	3	5.26	2	5.91
Anotia/microtia	18	2.12	5	1.28	9	2.64	0	0	4	7.01	0	0
Aortic valve stenosis	33	3.89	22	5.64	9	2.64	0	0	1	1.75	1	2.95
Atrial septal defect	113	13.31	42	10.77	55	16.15	3	11.06	6	10.52	7	20.68
Biliary atresia	6	0.71	3	0.77	1	0.29	0	0	2	3.51	0	0
Bladder exstrophy	2	0.24	0	0	2	0.59	0	0	0	0	0	0
Choanal atresia	20	2.36	7	1.79	11	3.23	0	0	1	1.75	1	2.95
Cleft lip with or without cleft palate	96	11.31	36	9.23	42	12.33	2	7.37	8	14.03	8	23.63
Cleft palate without cleft lip	53	6.25	29	7.43	15	4.4	0	0	8	14.03	1	2.95
Coarctation of aorta	37	4.36	19	4.87	13	3.82	1	3.69	2	3.51	2	5.91
Common truncus	6	0.71	1	0.26	5	1.47	0	0	0	0	0	0
Congenital cataract	13	1.53	4	1.03	7	2.06	1	3.69	1	1.75	0	0
Congenital hip dislocation	61	7.19	28	7.18	25	7.34	0	0	5	8.77	2	5.91
Diaphragmatic hernia	29	3.42	11	2.82	14	4.11	1	3.69	3	5.26	0	0
Down syndrome (Trisomy 21)	110	12.96	54	13.84	45	13.21	2	7.37	4	7.01	5	14.77
Ebstein's anomaly	9	1.06	4	1.03	4	1.17	0	0	1	1.75	0	0
Edwards syndrome (Trisomy 18)	25	2.95	13	3.33	7	2.06	2	7.37	2	3.51	1	2.95
Encephalocele	11	1.3	4	1.03	5	1.47	1	3.69	0	0	1	2.95
Endocardial cushion defect	31	3.65	13	3.33	13	3.82	0	0	3	5.26	1	2.95

[a] Incidence rates include live born and stillborn cases in numerator and only live borns in denominator.

[b] Incidence rates based on counts of less than 10 events are not statistically reliable.

[c] Total includes cases of unknown race/ethnicity, therefore it may be greater than the sum of the cases listed in the individual race/ethnicity categories.

[d] Many infants have multiple defects and are included in multiple condition categories. Therefore, the numbers in this row are NOT column totals.

Table 1 – C (continued)

Arizona Birth Defects Monitoring Program

Live-born and Stillborn Cases of Congenital Anomalies by Race/Ethnicity - Arizona 2000

Incidence Rate [a,b] Per 10,000 Live Births

CONDITION	TOTAL[c]	RATE	WHITE NON-HISP.	RATE	HISP.	RATE	BLACK	RATE	NATIVE AMER.	RATE	OTHER	RATE
Esophageal atresia / Tracheoesophageal fistula	18	2.12	9	2.31	8	2.35	0	0	1	1.75	0	0
Fetus or newborn affected by maternal alcohol use	3	0.35	0	0	0	0	0	0	2	3.51	0	0
Gastroschisis	46	5.42	21	5.38	16	4.7	3	11.06	4	7.01	1	2.95
Hirschsprung disease	12	1.41	5	1.28	5	1.47	0	0	0	0	2	5.91
Hydrocephalus without spina bifida	48	5.66	28	7.18	13	3.82	3	11.06	2	3.51	1	2.95
Hypoplastic left heart syndrome	15	1.77	7	1.79	6	1.76	1	3.69	1	1.75	0	0
Hypospadias and epispadias	221	26.04	144	36.92	52	15.27	6	22.12	8	14.03	10	29.54
Microcephaly	82	9.66	28	7.18	34	9.98	5	18.44	9	15.78	4	11.82
Obstructive genitourinary defect	151	17.79	69	17.69	62	18.2	7	25.81	10	17.53	3	8.86
Omphalocele	19	2.24	14	3.59	4	1.17	0	0	0	0	1	2.95
Patau syndrome (Trisomy 13)	16	1.89	7	1.79	7	2.06	0	0	0	0	2	5.91
Pulmonary valve atresia and stenosis	78	9.19	27	6.92	35	10.28	6	22.12	4	7.01	5	14.77
Pyloric stenosis	159	18.74	71	18.2	75	22.02	1	3.69	6	10.52	6	17.73
Rectal and large intestinal atresia/stenosis	33	3.89	15	3.85	14	4.11	2	7.37	1	1.75	1	2.95
Reduction deformity, lower limbs	22	2.59	7	1.79	13	3.82	0	0	2	3.51	0	0
Reduction deformity, upper limbs	37	4.36	20	5.13	14	4.11	3	11.06	0	0	0	0
Renal agenesis/hypoplasia	36	4.24	13	3.33	17	4.99	4	14.75	2	3.51	0	0
Spina bifida without anencephalus	29	3.42	13	3.33	13	3.82	0	0	2	3.51	0	0
Tetralogy of Fallot	32	3.77	20	5.13	9	2.64	0	0	2	3.51	1	2.95
Transposition of great arteries	42	4.95	15	3.85	17	4.99	0	0	5	8.77	3	8.86
Tricuspid valve atresia and stenosis	13	1.53	3	0.77	3	0.88	5	18.44	2	3.51	0	0
Ventricular septal defect	177	20.86	76	19.48	70	20.55	6	22.12	19	33.32	4	11.82

[a] Incidence rates include live born and stillborn cases in numerator and only live borns in denominator.

[b] Incidence rates based on counts of less than 10 events are not statistically reliable.

[c] Total includes cases of unknown race/ethnicity, therefore it may be greater than the sum of the cases listed in the individual race/ethnicity categories.

Table 2-A

Arizona Birth Defects Monitoring Program [a, b]

Birth Defects[c] by County of Residence, Arizona, 1998

STATE, COUNTY	LIVE BORNS (LB) WITH DEFECTS		STILLBORNS (SB) WITH DEFECTS		LIVE BORNS AND STILLBORNS WITH DEFECTS		NUMBER OF DEFECTS IN LIVE-BORN INFANTS		NUMBER OF DEFECTS IN STILLBORN INFANTS	
	Number	% OF ALL LB W/ DEFECTS	Number	% OF ALL SB W/ DEFECTS	Number	% OF BIRTHS W/ DEFECTS	Number	AVG # PER CHILD	Number	AVG # PER CHILD
ARIZONA	1390	1.78	52	8.4	1442	1.84	1816	1.31	83	1.6
APACHE COUNTY	24	1.79	0	0.00	24	1.78	38	1.58	0	0.00
COCHISE COUNTY	26	1.59	3	20.00	29	1.76	30	1.15	5	1.67
COCONINO COUNTY	25	1.42	0	0.00	25	1.41	38	1.52	0	0.00
GILA COUNTY	9	1.27	1	20.00	10	1.40	11	1.22	0	0.00
GRAHAM COUNTY	6	1.23	0	0.00	6	1.22	8	1.33	0	0.00
GREENLEE COUNTY	3	2.11	1	33.33	4	2.76	3	1.00	1	1.00
LA PAZ COUNTY	4	2.29	0	0.00	4	2.29	4	1.00	0	0.00
MARICOPA COUNTY	886	1.80	31	8.09	917	1.85	1131	1.28	54	1.74
MOHAVE COUNTY	20	1.13	1	6.25	21	1.18	25	1.25	1	1.00
NAVAJO COUNTY	37	2.09	2	11.11	39	2.18	43	1.16	2	1.00
PIMA COUNTY	214	1.87	6	6.00	220	1.91	299	1.40	9	1.50
PINAL COUNTY	43	1.92	1	5.26	44	1.95	60	1.40	2	2.00
SANTA CRUZ COUNTY	12	1.55	0	0.00	12	1.53	12	1.00	0	0.00
YAVAPAI COUNTY	36	2.12	5	31.25	41	2.40	52	1.44	8	1.60
YUMA COUNTY	45	1.60	1	9.09	46	1.63	62	1.38	1	1.00

[a]Total number of live births in Arizona for 1998 = 77,945. [b]Total number of fetal deaths in Arizona for 1998 = 619. [c]Includes all 44 birth defect categories monitored. See Appendix B.

Table 2-B

Arizona Birth Defects Monitoring Program [a, b]

Birth Defects[c] by County of Residence, Arizona,1999

STATE, COUNTY	LIVE BORNS (LB) WITH DEFECTS		STILLBORNS (SB) WITH DEFECTS		LIVE BORNS AND STILLBORNS WITH DEFECTS		NUMBER OF DEFECTS IN LIVE-BORN INFANTS		NUMBER OF DEFECTS IN STILLBORN INFANTS	
	Number	% OF ALL LB W/ DEFECTS	Number	% OF ALL SB W/ DEFECTS	Number	% OF BIRTHS W/ DEFECTS	Number	AVG # PER CHILD	Number	AVG # PER CHILD
ARIZONA	1423	1.76	51	8.43	1474	1.81	1832	1.29	77	1.51
APACHE COUNTY	26	2.00	0	0.00	26	1.99	30	1.15	0	0.00
COCHISE COUNTY	32	1.92	2	15.38	34	2.02	41	1.28	6	3.00
COCONINO COUNTY	38	2.09	1	7.14	39	2.13	57	1.50	1	1.00
GILA COUNTY	16	2.39	0	0.00	16	2.37	27	1.69	0	0.00
GRAHAM COUNTY	13	2.70	0	0.00	13	2.68	13	1.00	0	0.00
GREENLEE COUNTY	3	2.10	0	0.00	3	2.08	3	1.00	0	0.00
LA PAZ COUNTY	2	1.14	0	0.00	2	1.12	2	1.00	0	0.00
MARICOPA COUNTY	887	1.72	33	8.40	920	1.77	1149	1.30	46	1.39
MOHAVE COUNTY	26	1.38	0	0.00	26	1.37	32	1.23	0	0.00
NAVAJO COUNTY	30	1.69	0	0.00	30	1.68	38	1.27	0	0.00
PIMA COUNTY	220	1.86	10	11.49	230	1.93	279	1.27	13	1.30
PINAL COUNTY	44	1.83	0	0.00	44	1.81	59	1.34	0	0.00
SANTA CRUZ COUNTY	17	2.25	1	16.67	18	2.36	22	1.29	1	1.00
YAVAPAI COUNTY	23	1.42	1	14.29	24	1.48	28	1.22	2	2.00
YUMA COUNTY	46	1.61	3	15.79	49	1.70	52	1.13	8	2.67

[a]Total number of live births in Arizona for 1999 = 80,908. [b]Total number of fetal deaths in Arizona for 1999 = 605 [c]Includes all 44 birth defect categories monitored. See Appendix B.

Table 2-C

Arizona Birth Defects Monitoring Program [a, b]

Birth Defects[c] by County of Residence, Arizona, 2000

STATE, COUNTY	LIVE BORNS (LB) WITH DEFECTS		STILLBORNS (SB) WITH DEFECTS		LIVE BORNS AND STILLBORNS WITH DEFECTS		NUMBER OF DEFECTS IN LIVE-BORN INFANTS		NUMBER OF DEFECTS IN STILLBORN INFANTS	
	Number	% OF ALL LB W/ DEFECTS	Number	% OF ALL SB W/ DEFECTS	Number	% OF BIRTHS W/ DEFECTS	Number	AVG # PER CHILD	Number	AVG # PER CHILD
ARIZONA	1526	1.79	74	11.18	1600	1.87	1975	1.29	103	1.39
APACHE COUNTY	18	1.36	0	0.00	18	1.35	31	1.72	0	0.00
COCHISE COUNTY	25	1.44	4	30.77	29	1.65	30	1.20	4	1.00
COCONINO COUNTY	38	2.04	2	16.67	40	2.13	54	1.42	4	2.00
GILA COUNTY	12	1.79	1	10.00	13	1.91	18	1.50	1	1.00
GRAHAM COUNTY	12	2.56	0	0.00	12	2.55	17	1.42	0	0.00
GREENLEE COUNTY	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
LA PAZ COUNTY	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00
MARICOPA COUNTY	983	1.81	44	10.65	1027	1.87	1255	1.28	59	1.34
MOHAVE COUNTY	34	1.92	0	0.00	34	1.91	50	1.47	0	0.00
NAVAJO COUNTY	19	1.13	0	0.00	19	1.12	29	1.53	0	0.00
PIMA COUNTY	242	1.94	16	13.22	258	2.05	296	1.22	27	1.69
PINAL COUNTY	56	2.15	2	8.70	58	2.20	72	1.29	4	2.00
SANTA CRUZ COUNTY	13	1.63	0	0.00	13	1.61	17	1.31	0	0.00
YAVAPAI COUNTY	30	1.71	1	9.09	31	1.76	49	1.63	0	0.00
YUMA COUNTY	44	1.46	4	18.18	48	1.59	57	1.30	4	1.00

[a]Total number of live births in Arizona for 2000 = 84,866. [b]Total number of fetal deaths in Arizona for 2000 = 662. [c]Includes all 44 birth defect categories monitored. See Appendix B.

Table 3

Congenital Anomalies[a] by Year, Live Borns and Stillborns, 1986 - 2000

Incidence Rates Per 10,000 Live Births, Arizona

CONDITION		1986	1987	1988	1989	1990	1991	1992	1993	1994	1995	1996	1997	1998	1999	2000
Amniotic bands	Cases[b]	4	4	9	8	14	10	8	8	9	12	6	13	14	10	14
	Rate	0.67	0.64	1.39	1.19	2.04	1.47	1.17	1.16	1.27	1.66	0.8	1.72	1.8	1.24	1.65
Anencephaly	Cases	21	17	17	19	16	18	21	15	29	18	23	25	19	15	14
	Rate	3.51	2.73	2.63	2.83	2.33	2.65	3.06	2.17	4.09	2.49	3.06	3.31	2.44	1.85	1.65
Aniridia	Cases	0	1	0	0	2	0	2	0	0	2	1	2	3	0	1
	Rate	0	0.16	0	0	0.29	0	0.29	0	0	0.28	0.13	0.26	0.38	0	0.12
Anophthalmia/microphthalmia	Cases	15	25	24	23	30	34	24	16	18	25	20	19	13	14	22
	Rate	2.5	4.01	3.72	3.43	4.36	5	3.5	2.32	2.54	3.45	2.66	2.51	1.67	1.73	2.59
Anotia/microtia	Cases	23	42	20	18	17	31	20	13	11	25	21	31	22	28	18
	Rate	3.84	6.73	3.1	2.68	2.47	4.56	2.91	1.88	1.55	3.45	2.8	4.1	2.82	3.46	2.12
Aortic valve stenosis	Cases	8	15	17	25	17	17	22	15	21	30	19	27	26	42	33
	Rate	1.34	2.41	2.63	3.73	2.47	2.5	3.21	2.17	2.96	4.14	2.53	3.57	3.34	5.19	3.89
Atrial septal defect	Cases	44	90	95	131	111	116	80	85	70	83	74	99	122	106	113
	Rate	7.35	14.43	14.71	19.54	16.15	17.06	11.66	12.32	9.87	11.46	9.86	13.1	15.65	13.1	13.31
Biliary atresia	Cases	2	1	3	5	4	6	4	8	6	3	7	2	5	4	6
	Rate	0.33	0.16	0.46	0.75	0.58	0.88	0.58	1.16	0.85	0.41	0.93	0.26	0.64	0.49	0.71
Bladder exstrophy	Cases	1	3	1	1	2	2	4	1	1	3	1	1	2	2	2
	Rate	0.17	0.48	0.15	0.15	0.29	0.29	0.58	0.14	0.14	0.41	0.13	0.13	0.26	0.25	0.24
Choanal atresia	Cases	6	10	10	16	6	5	6	7	10	14	18	11	12	14	20
	Rate	1	1.6	1.55	2.39	0.87	0.74	0.87	1.01	1.41	1.93	2.4	1.46	1.54	1.73	2.36
Cleft lip with or without cleft palate	Cases	77	80	91	90	97	80	74	92	83	93	87	103	95	90	96
	Rate	12.86	12.83	14.09	13.42	14.11	11.77	10.78	13.33	11.71	12.84	11.59	13.62	12.19	11.12	11.31
Cleft palate without cleft lip	Cases	43	48	36	48	45	37	31	53	52	49	47	44	53	58	53
	Rate	7.18	7.7	5.57	7.16	6.55	5.44	4.52	7.68	7.34	6.77	6.26	5.82	6.8	7.17	6.25
Coarctation of aorta	Cases[b]	24	26	41	40	29	25	30	24	18	40	39	38	37	41	37
	Rate	1.5	2.57	1.24	2.39	2.76	1.62	1.89	2.03	1.27	1.38	2	1.59	1.03	1.61	1.77
Common truncus	Cases	4	10	9	9	6	6	3	4	6	3	7	11	11	1	6
	Rate	0.67	1.6	1.39	1.34	0.87	0.88	0.44	0.58	0.85	0.41	0.93	1.46	1.41	0.12	0.71
Congenital cataract	Cases	8	7	7	15	24	10	12	8	14	14	9	13	11	14	13
	Rate	1.34	1.12	1.08	2.24	3.49	1.47	1.75	1.16	1.97	1.93	1.2	1.72	1.41	1.73	1.53
Congenital hip dislocation	Cases	87	101	68	91	105	103	66	74	65	83	67	55	63	57	61
	Rate	14.53	16.19	10.53	13.57	15.27	15.15	9.62	10.73	9.17	11.46	8.92	7.28	8.08	7.05	7.19

[a] See Appendix A and Appendix B for definitions of the conditions. [b]"Cases" is the number of live born and stillborn infants >= 20 weeks gestation.

Table 3 (continued)

Congenital Anomalies[a] by Year, Live Borns and Stillborns, 1986 - 2000

Incidence Rates Per 10,000 Live Births, Arizona

CONDITION		1986	1987	1988	1989	1990	1991	1992	1993	1994	1995	1996	1997	1998	1999	2000
Diaphragmatic hernia	Cases[b]	13	18	20	23	28	23	13	18	21	20	15	15	30	19	29
	Rate	2.17	2.89	3.1	3.43	4.07	3.38	1.89	2.61	2.96	2.76	2	1.98	3.85	2.35	3.42
Down syndrome (Trisomy 21)	Cases	64	62	74	66	73	83	88	81	100	90	96	101	125	115	110
	Rate	10.69	9.94	11.46	9.84	10.62	12.21	12.82	11.74	14.11	12.43	12.79	13.36	16.04	14.21	12.96
Ebstein's anomaly	Cases	5	3	6	7	5	10	7	8	7	6	11	9	5	8	9
	Rate	0.83	0.48	0.93	1.04	0.73	1.47	1.02	1.16	0.99	0.83	1.46	1.19	0.64	0.99	1.06
Edwards syndrome (Trisomy 18)	Cases	11	17	13	10	15	13	12	14	14	18	16	15	12	18	25
	Rate	1.84	2.73	2.01	1.49	2.18	1.91	1.75	2.03	1.97	2.49	2.13	1.98	1.54	2.22	2.95
Encephalocele	Cases	9	8	14	5	13	14	2	6	11	6	15	9	8	11	11
	Rate	1.5	1.28	2.17	0.75	1.89	2.06	0.29	0.87	1.55	0.83	2	1.19	1.03	1.36	1.3
Endocardial cushion defect	Cases	15	22	30	25	29	25	24	28	28	34	27	29	44	37	31
	Rate	2.5	3.53	4.65	3.73	4.22	3.68	3.5	4.06	3.95	4.7	3.6	3.84	5.65	4.57	3.65
Esophageal atresia/tracheoesophageal fistula	Cases	19	16	19	18	19	16	14	13	14	18	16	22	19	14	18
	Rate	3.17	2.57	2.94	2.68	2.76	2.35	2.04	1.88	1.97	2.49	2.13	2.91	2.44	1.73	2.12
Fetus or newborn affected by maternal alcohol use	Cases	9	25	12	21	22	27	33	26	14	27	10	9	6	12	3
	Rate	1.5	4.01	1.86	3.13	3.2	3.97	4.81	3.77	1.97	3.73	1.33	1.19	0.77	1.48	0.35
Gastroschisis	Cases	18	18	19	19	20	36	27	15	27	27	42	36	39	28	46
	Rate	3.01	2.89	2.94	2.83	2.91	5.3	3.93	2.17	3.81	3.73	5.59	4.76	5	3.46	5.42
Hirschsprung disease	Cases	12	10	10	7	13	13	7	8	10	16	10	8	8	13	12
	Rate	2	1.6	1.55	1.04	1.89	1.91	1.02	1.16	1.41	2.21	1.33	1.06	1.03	1.61	1.41
Hydrocephalus without spina bifida	Cases	36	44	48	46	51	50	38	30	43	42	39	32	44	44	48
	Rate	6.01	7.05	7.43	6.86	7.42	7.35	5.54	4.35	6.07	5.8	5.19	4.23	5.65	5.44	5.66
Hypoplastic left heart syndrome	Cases	9	16	8	16	19	11	13	14	9	10	15	12	8	13	15
	Rate	1.5	2.57	1.24	2.39	2.76	1.62	1.89	2.03	1.27	1.38	2	1.59	1.03	1.61	1.77
Hypospadias and epispadias	Cases	161	160	157	160	176	160	162	133	142	181	197	201	228	234	221
	Rate	26.88	25.65	24.31	23.86	25.6	23.54	23.6	19.28	20.03	25	26.24	26.59	29.25	28.92	26.04
Microcephaly	Cases	30	60	70	109	118	120	90	84	75	81	67	69	53	70	82
	Rate	5.01	9.62	10.84	16.26	17.16	17.65	13.11	12.18	10.58	11.19	8.92	9.13	6.8	8.65	9.66
Obstructive genitourinary defect	Cases	44	75	64	91	91	105	80	83	92	109	121	112	125	122	151
	Rate	7.35	12.03	9.91	13.57	13.24	15.45	11.66	12.03	12.98	15.05	16.11	14.81	16.04	15.08	17.79
Omphalocele	Cases	10	14	17	10	21	21	10	17	11	14	21	10	14	14	19
	Rate	1.67	2.24	2.63	1.49	3.05	3.09	1.46	2.46	1.55	1.93	2.8	1.32	1.8	1.73	2.24

[a] See Appendix A and Appendix B for definitions of the conditions. [b]"Cases" is the number of live born and stillborn infants >= 20 weeks gestation.

Table 3 (continued)

Congenital Anomalies[a] by Year, Live Borns and Stillborns, 1986 - 2000

Incidence Rates Per 10,000 Live Births, Arizona

CONDITION		1986	1987	1988	1989	1990	1991	1992	1993	1994	1995	1996	1997	1998	1999	2000
Patau Syndrome (Trisomy 13)	Cases	8	4	3	4	11	6	15	9	3	8	12	5	10	8	16
	Rate	1.34	0.64	0.46	0.6	1.6	0.88	2.19	1.3	0.42	1.1	1.6	0.66	1.28	0.99	1.89
Pulmonary valve atresia and stenosis	Cases	32	44	55	61	54	55	56	50	60	69	63	67	56	69	78
	Rate	5.34	7.05	8.52	9.1	7.85	8.09	8.16	7.25	8.46	9.53	8.39	8.86	7.18	8.53	9.19
Pyloric stenosis	Cases	108	135	134	122	116	148	137	127	159	148	140	152	135	145	159
	Rate	18.03	21.65	20.75	18.2	16.87	21.77	19.96	18.41	22.43	20.44	18.65	20.11	17.32	17.92	18.74
Rectal and large intestinal atresia/stenosis	Cases	29	30	31	36	36	39	33	30	28	42	30	20	43	28	33
	Rate	4.84	4.81	4.8	5.37	5.24	5.74	4.81	4.35	3.95	5.8	4	2.65	5.52	3.46	3.89
Reduction deformity, lower limbs	Cases	13	17	22	25	34	15	12	17	13	14	14	20	20	17	22
	Rate	2.17	2.73	3.41	3.73	4.95	2.21	1.75	2.46	1.83	1.93	1.86	2.65	2.57	2.1	2.59
Reduction deformity, upper limbs	Cases	39	42	51	50	61	41	25	25	30	32	23	28	23	29	37
	Rate	6.51	6.73	7.9	7.46	8.87	6.03	3.64	3.62	4.23	4.42	3.06	3.7	2.95	3.58	4.36
Renal agenesis/hypoplasia	Cases	21	27	22	42	33	37	33	30	25	39	39	34	36	30	36
	Rate	3.51	4.33	3.41	6.26	4.8	5.44	4.81	4.35	3.53	5.39	5.19	4.5	4.62	3.71	4.24
Spina bifida without anencephalus	Cases	33	34	24	38	37	31	36	35	32	33	32	37	33	40	29
	Rate	5.51	5.45	3.72	5.67	5.38	4.56	5.25	5.07	4.51	4.56	4.26	4.89	4.23	4.94	3.42
Tetralogy of Fallot	Cases	15	18	29	23	27	22	32	30	30	29	34	33	33	25	32
	Rate	2.5	2.89	4.49	3.43	3.93	3.24	4.66	4.35	4.23	4.01	4.53	4.37	4.23	3.09	3.77
Transposition of great arteries	Cases	33	27	26	33	28	26	25	28	30	33	34	40	36	39	42
	Rate	5.51	4.33	4.03	4.92	4.07	3.82	3.64	4.06	4.23	4.56	4.53	5.29	4.62	4.82	4.95
Tricuspid valve atresia and stenosis	Cases	14	16	11	8	6	10	7	7	13	10	7	8	7	6	13
	Rate	2.34	2.57	1.7	1.19	0.87	1.47	1.02	1.01	1.83	1.38	0.93	1.06	0.9	0.74	1.53
Ventricular septal defect	Cases	94	131	100	135	132	128	119	117	115	141	146	145	127	143	177
	Rate	15.69	21	15.48	20.13	19.2	18.83	17.34	16.96	16.22	19.47	19.44	19.18	16.29	17.67	20.86

[a] See Appendix A and Appendix B for definitions of the conditions. [b]"Cases" is the number of live born and stillborn infants >= 20 weeks gestation.

The rates are calculated as the number of live born and stillborn cases of each defect in each year divided by the total number of live births in each year.
Live births for each year are as follows:

Year	1986	1987	1988	1989	1990	1991	1992	1993	1994	1995	1996	1997	1998	1999	2000
# of live births in Arizona to Arizona residents	59,892	62,370	64,584	67,050	68,751	67,981	68,635	68,993	70,892	72,404	75,087	75,601	77,945	80,908	84,867

Figure 1:

Trends of Selected Congenital Anomalies, Incidence Rates, 1986-2000 (Live Born and Stillborn Cases per 10,000 Live Births)

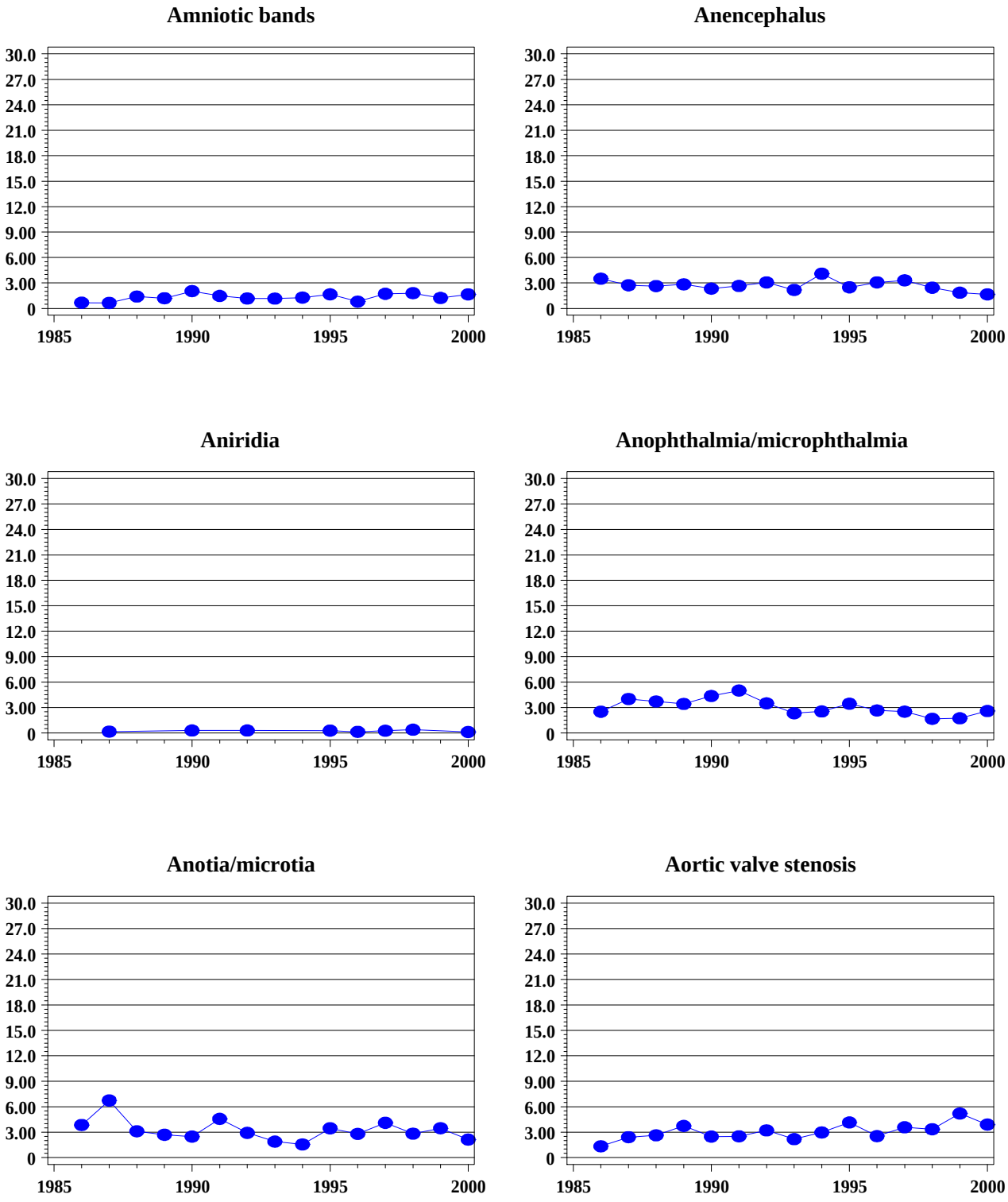


Figure 1:

Trends of Selected Congenital Anomalies, Incidence Rates, 1986-2000 (Live Born and Stillborn Cases per 10,000 Live Births)

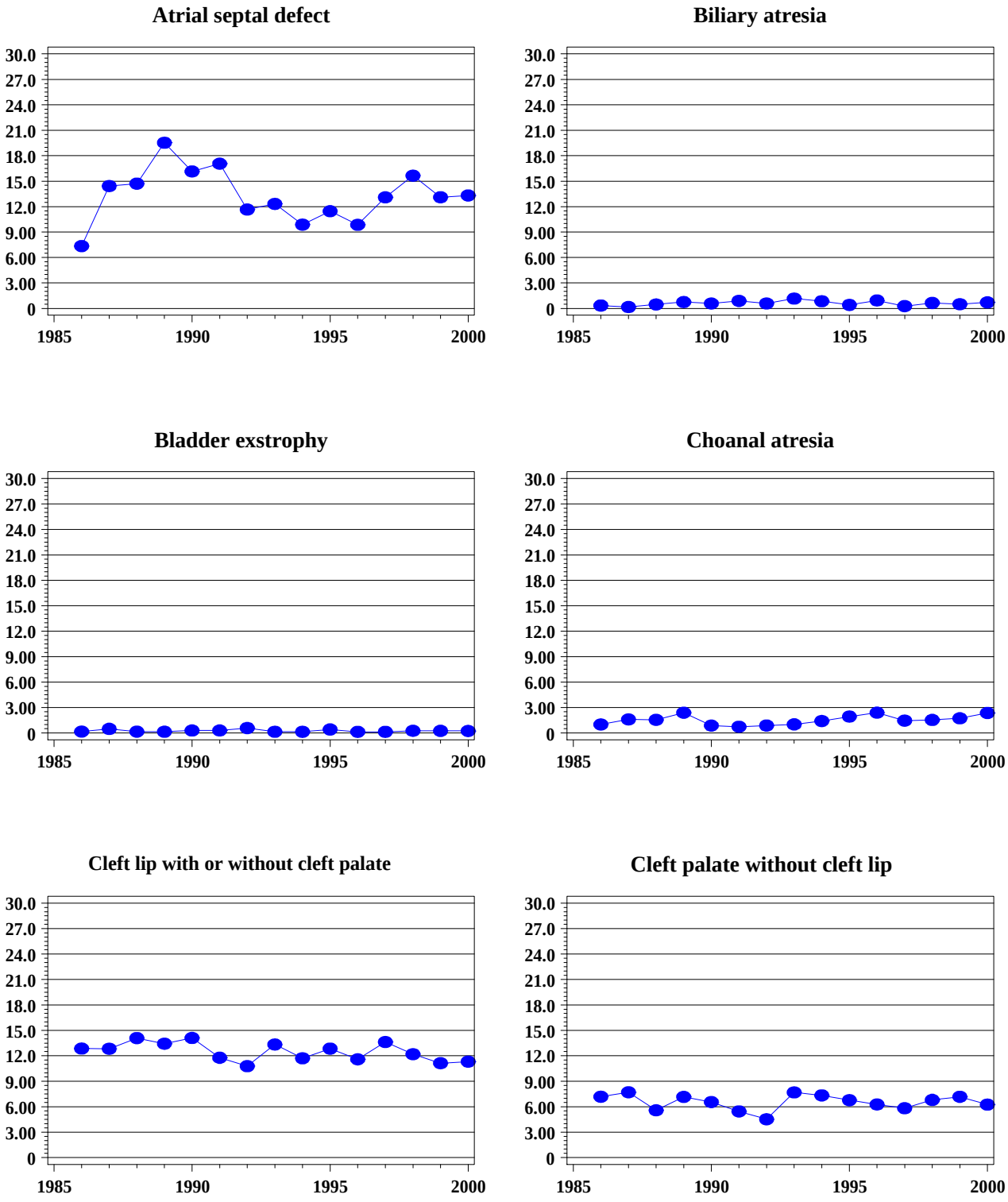


Figure 1:

Trends of Selected Congenital Anomalies, Incidence Rates, 1986-2000 (Live Born and Stillborn Cases per 10,000 Live Births)

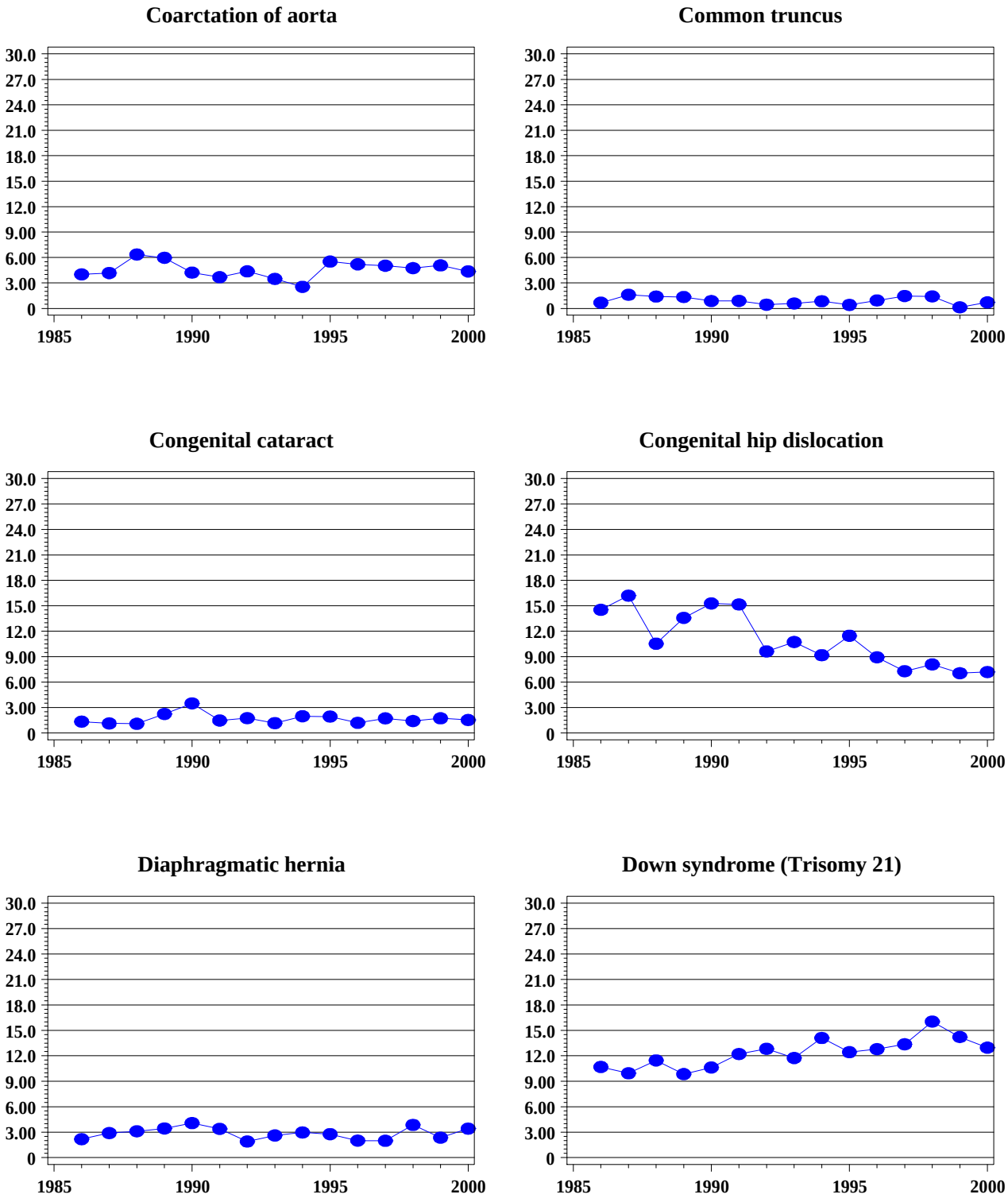
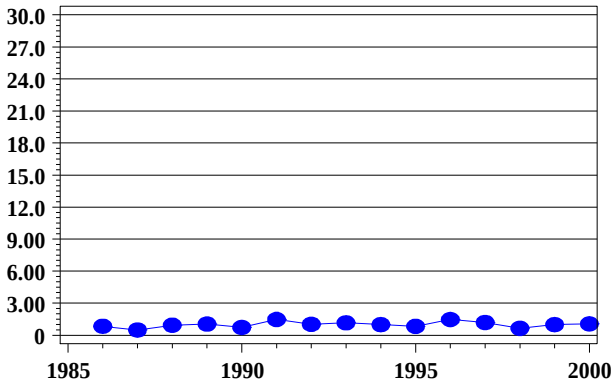


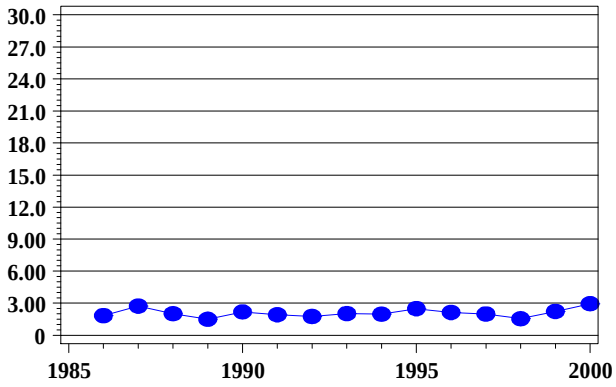
Figure 1:

Trends of Selected Congenital Anomalies, Incidence Rates, 1986-2000 (Live Born and Stillborn Cases per 10,000 Live Births)

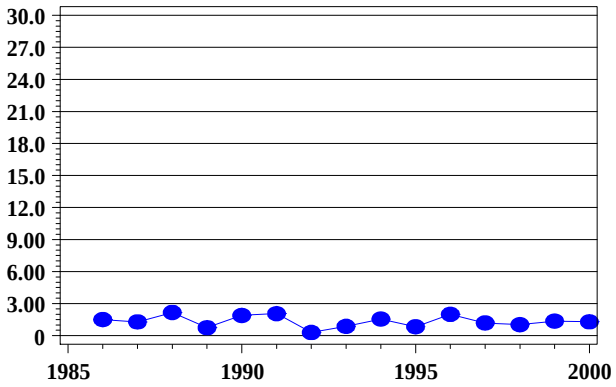
Ebstein anomaly



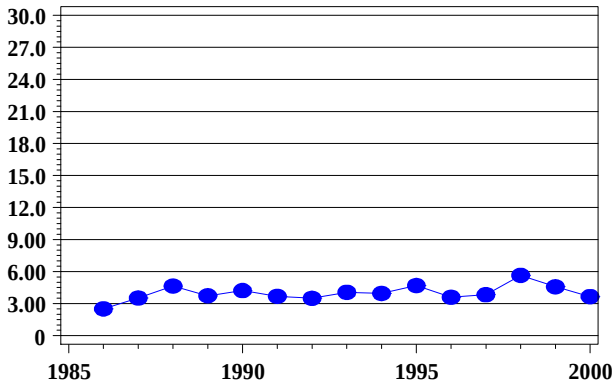
Edwards syndrome (Trisomy 18)



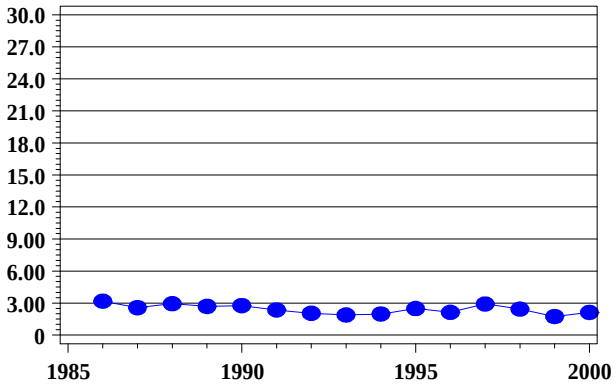
Encephalocele



Endocardial cushion defect



Esophageal atresia/TE fistula



Fetal alcohol syndrome

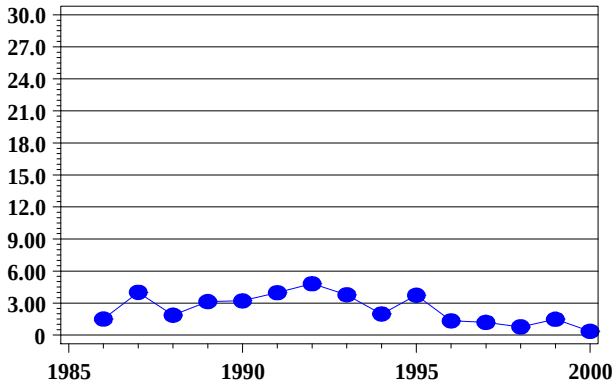
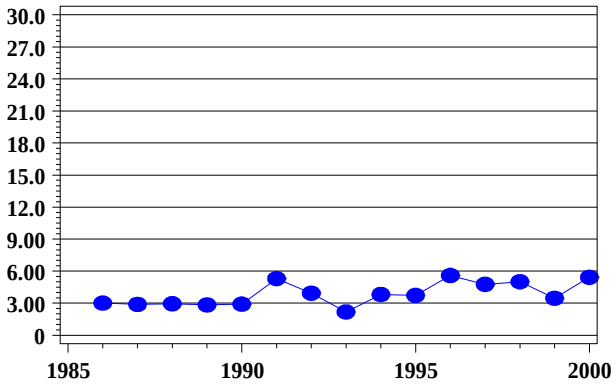


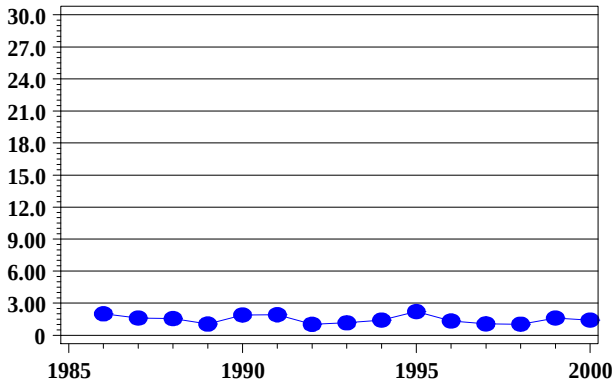
Figure 1:

Trends of Selected Congenital Anomalies, Incidence Rates, 1986-2000 (Live Born and Stillborn Cases per 10,000 Live Births)

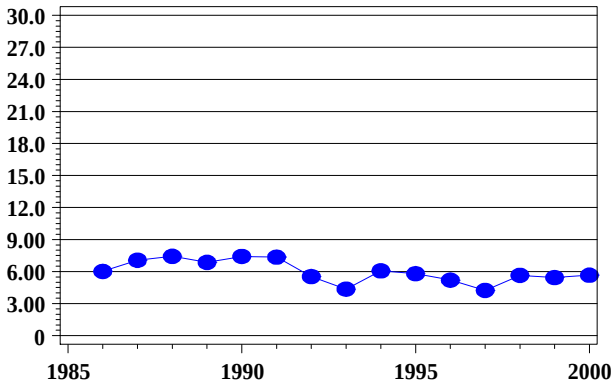
Gastroschisis



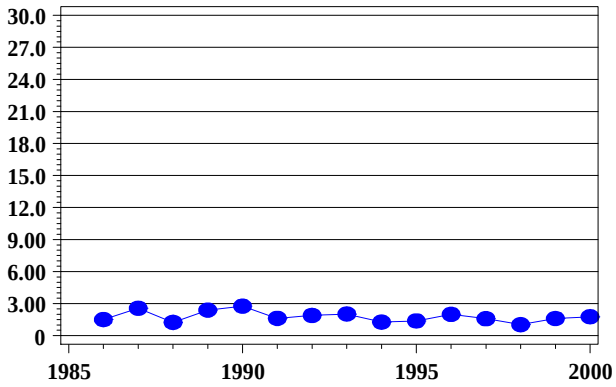
Hirschsprung disease



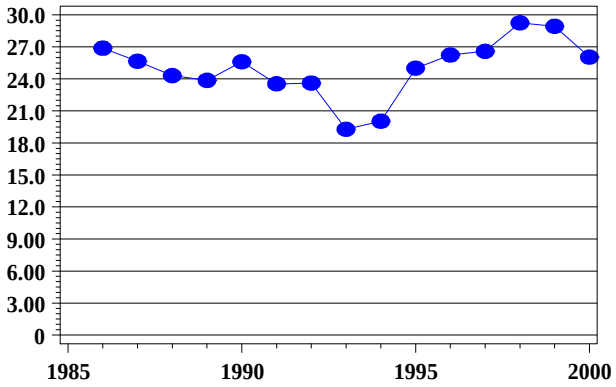
Hydrocephalus without spina bifida



Hypoplastic left heart syndrome



Hypospadias and Epispadias



Microcephalus

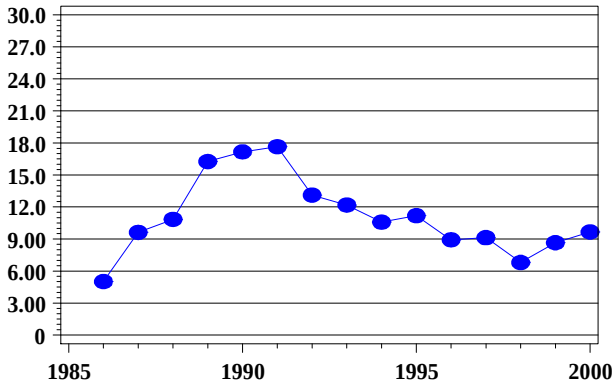
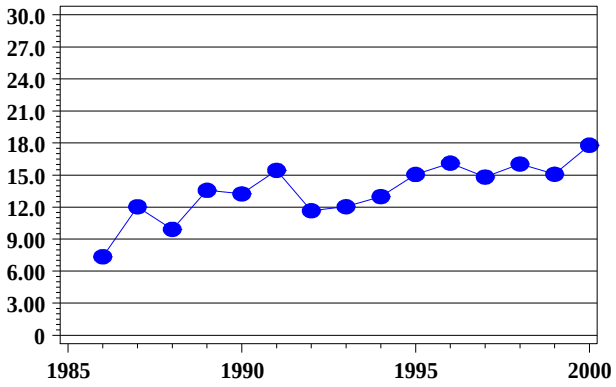


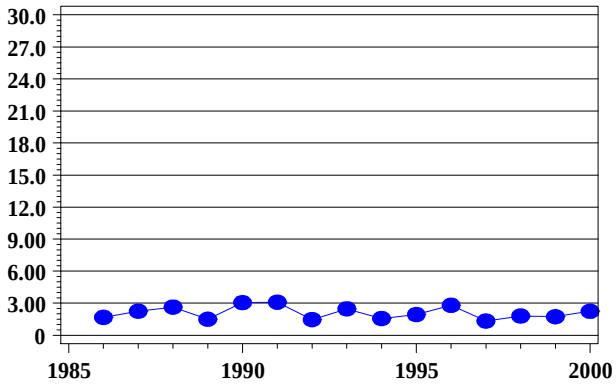
Figure 1:

Trends of Selected Congenital Anomalies, Incidence Rates, 1986-2000 (Live Born and Stillborn Cases per 10,000 Live Births)

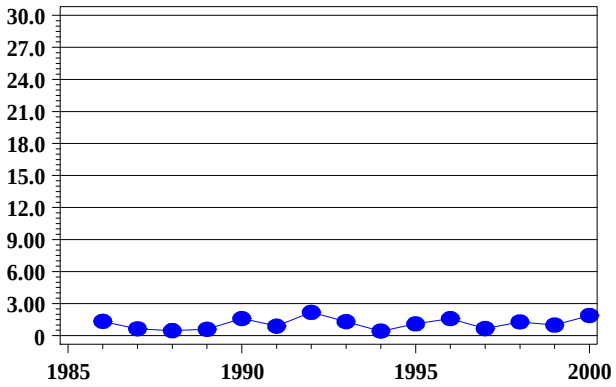
Obstructive genitourinary defect



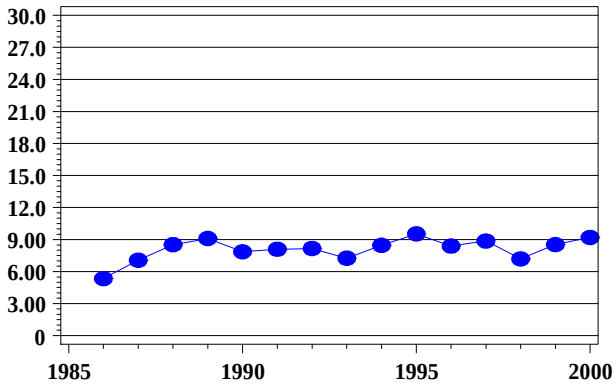
Omphalocele



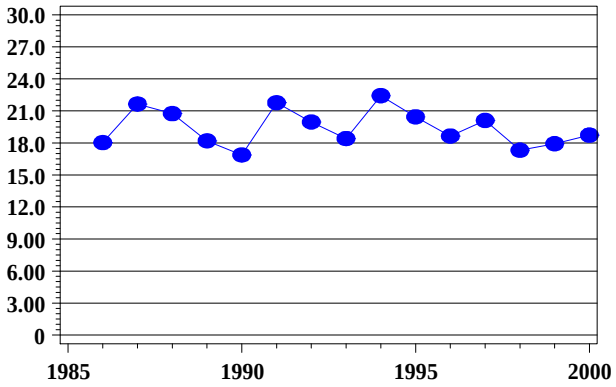
Patau syndrome (Trisomy 13)



Pulmonary valve atresia and stenosis



Pyloric stenosis



Rectal/large intestinal atresia/stenosis

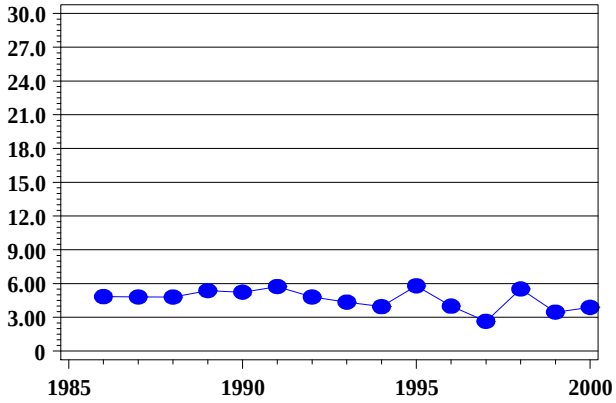
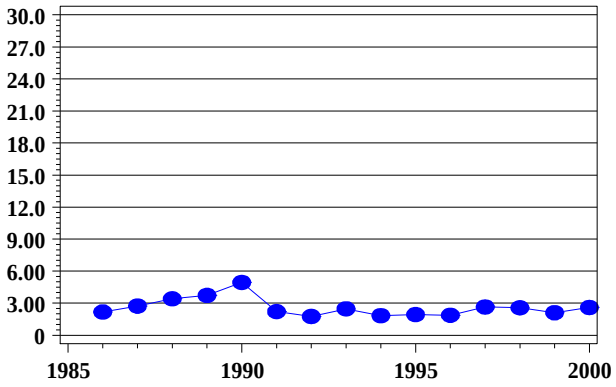


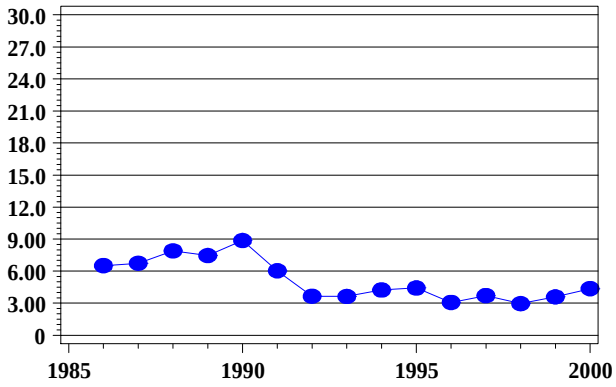
Figure 1:

Trends of Selected Congenital Anomalies, Incidence Rates, 1986-2000 (Live Born and Stillborn Cases per 10,000 Live Births)

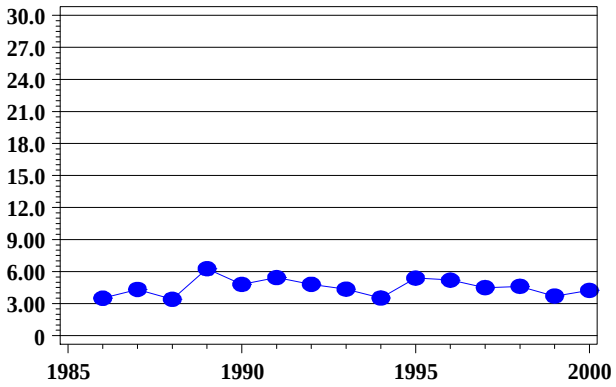
Reduction deformity, lower limbs



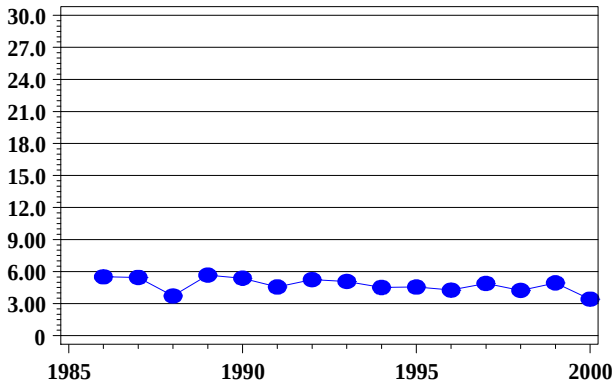
Reduction deformity, upper limbs



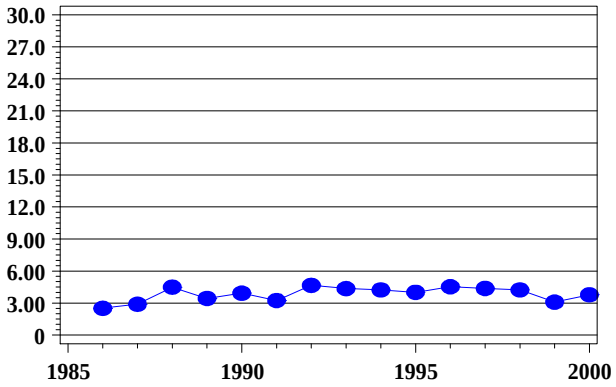
Renal agenesis/hypoplasia



Spina bifida without anencephalus



Tetralogy of Fallot



Transposition of great arteries

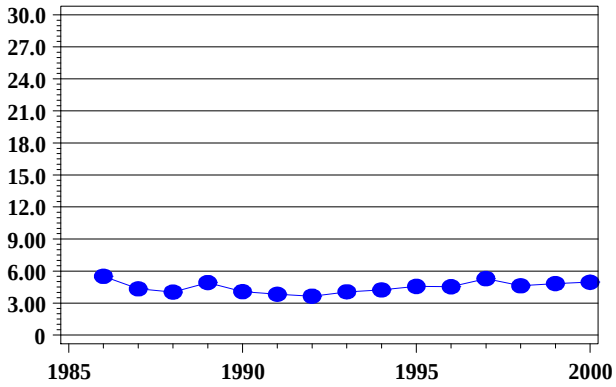
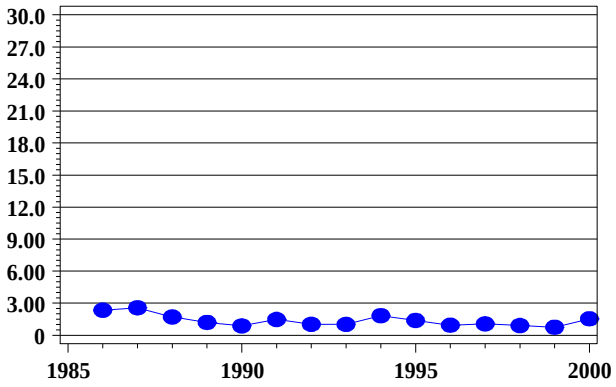


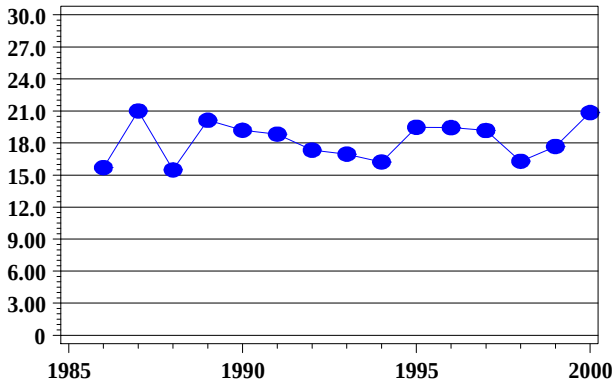
Figure 1:

Trends of Selected Congenital Anomalies, Incidence Rates, 1986-2000 (Live Born and Stillborn Cases per 10,000 Live Births)

Tricuspid valve atresia and stenosis

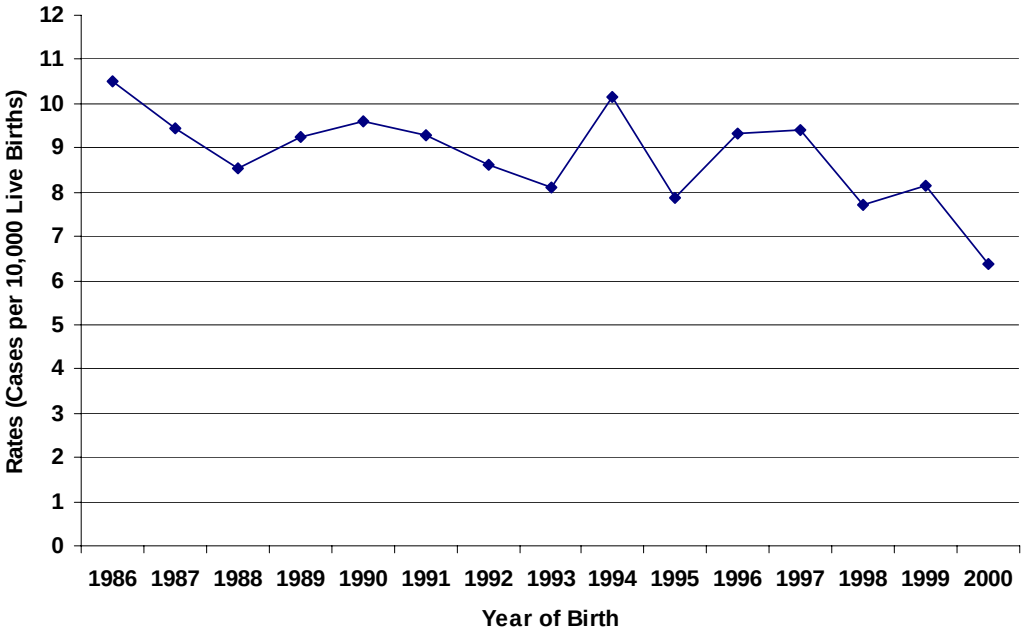


Ventricular septal defect



NEURAL TUBE DEFECTS

Figure 2: Neural Tube Defect Incidence Rates, Arizona, 1986-2000



Neural tube defects (NTDs) result from the failure of the neural tube to close properly in fetal development, occurring at approximately four weeks gestation. The three major NTDs are anencephaly, encephalocele, and spina bifida. Anencephaly is an absence of part or all of the brain. Encephalocele is the herniation of brain tissue through a gap in the skull. Spina bifida is a defective closure of the bones of the spine, through which the spinal cord and meninges may or may not protrude. Research indicates that maternal obesity, socioeconomic status and neighborhood social conditions, prior spontaneous and elective terminations, and short periods of time between pregnancies are associated with an increased risk for an NTD-affected pregnancy.[25,26,27] However, compelling data from randomized clinical trials shows that daily intake of 400 mcg of folic acid starting before conception and continuing through the first trimester can reduce the risk of an NTD-affected pregnancy by at least 50 percent.[28,29]

Because of the overwhelming evidence that folic acid helps to prevent these birth defects, the Food and Drug Administration mandated fortification of cereal grain products with 140 mcg/100 g starting in January of 1998.[30] Since then, there has been a 26% decrease in NTD-

affected pregnancies in the United States.[31] The pre-fortification NTD rate in Arizona (1995 and 1996 data combined) was 8.61 cases per 10,000 live births. The post-fortification NTD rate in Arizona (1999 and 2000 data combined) was 7.24 cases per 10,000 live births. This shows a 16% decline in NTD rates in Arizona post-fortification. It is possible that the decline in NTDs in Arizona is less than the decline in the United States because Hispanics (39% of women delivering live born infants in the year 2000 in Arizona were Hispanic) traditionally eat fewer wheat products (most of which are fortified) and more corn flour products (most of which are not fortified) than other ethnic groups in the country. Because NTD rates are highest among infants born to Hispanic mothers across the country, the Spina Bifida Association, the National Council of La Raza, Wal-Mart Stores, Inc., and GRUMA S.A. de C.V., internationally one of the largest producers of corn flour and corn flour products, formed a partnership as of July of 2006 to encourage corn meal suppliers in North America to start fortifying their corn flour products with folic acid.[32]

RACE/ETHNICITY

All race and ethnic groups experience birth defects, but the frequency and types of these defects vary by race and ethnicity.[33,34] The race and ethnicity information collected in the Arizona birth and fetal death certificates allow for the analysis of birth defects by race and ethnicity. See Appendix E for an explanation of how race/ethnicity is determined for each case.

Figure 3 displays the rates of spina bifida without anencephalus by race and ethnicity for births for 1998 to 2000, combined. (See Tables 1-A, 1-B, and 1-C for data.) The overall rate of spina bifida without anencephaly for Hispanics and Whites was significantly higher than it was for Blacks. Arizona data in 1998-2000 also shows that the rate of spina bifida without anencephaly was higher in Hispanics than it was in Whites. However, the difference between Hispanic and White Non-Hispanic incidence rates for 1998-2000 in Arizona was not statistically significant.

Figure 3. Spina Bifida without Anencephalus, Incidence Rates (Live Born and Stillborn Cases Per 10,000 Live Births) by Race/Ethnicity, Arizona, 1998-2000

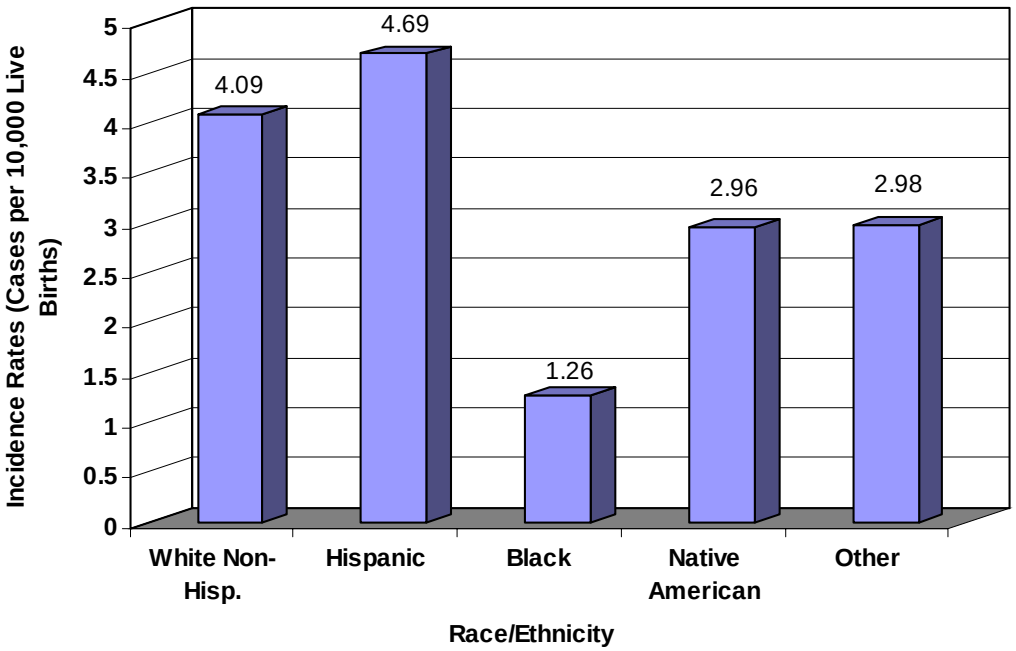
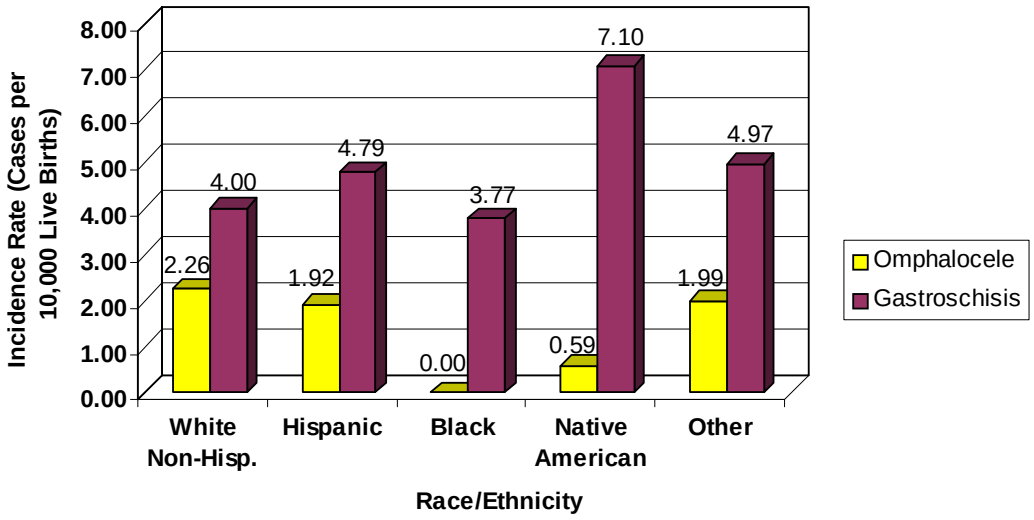


Figure 4 shows the rates of gastroschisis and omphalocele for 1998 to 2000. The rate of gastroschisis among Native Americans was significantly higher than the rates for Whites and Hispanics (7.10 for Native American v. 4.00 for Whites and 4.79 for Hispanics per 10,000 live births). In contrast, the rate of omphalocele in Native Americans was significantly lower than the rates for Whites and Hispanics (0.59 for Native American v. 2.26 for Whites and 1.92 for Hispanics per 10,000 live and still births).

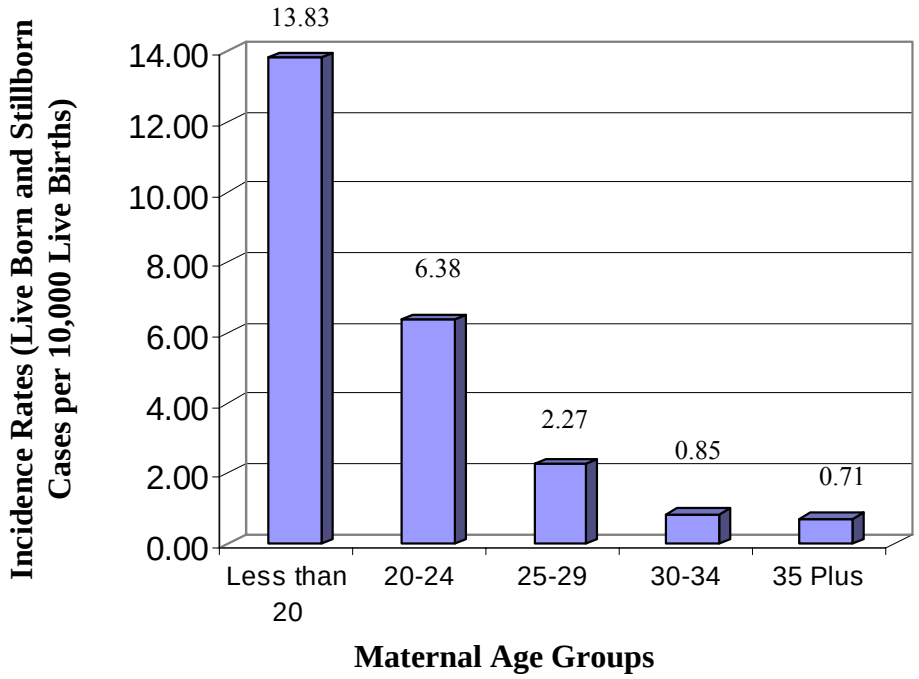
Figure 4. Abdominal Wall Defect Incidence Rates (Live Born and Stillborn Cases Per 10,000 Live Births) by Race/Ethnicity, Arizona, 1998-2000



MATERNAL AGE

Maternal age was divided into five age groups: less than 20 years of age, 20 to 24 years of age, 25 to 29 years of age, 30 to 34 years of age, and 35 years of age and older. Figure 5 demonstrates that gastroschisis incidence rates for 1998-2000 decrease with increasing maternal age. The rates of gastroschisis for the less than 20 years of age and the 20 to 24 years of age maternal age groups were statistically higher than the state rate for gastroschisis. It has been reported in the literature that teen mothers are six times more likely as women 25 years of age or older to have a child with gastroschisis.[35] In Arizona from 1998-2000, teen mothers were 3.6 times as likely as women 25 years or older to have a child with gastroschisis.

Figure 5: Gastroschisis Incidence Rates, Arizona, 1998-2000
(Live Born and Still Born Cases Per 10,000 Live Births, by Maternal Age Groups)



COUNTY PATTERNS FOR SENTINEL DEFECTS

Tables 4-7 look at the number of cases and incidence rates statewide and by county for 1986-2000 data combined for the following sentinel defects: neural tube defects, gastroschisis, omphalocele, and heart defects. These defects were chosen because of their significant public health impact.

Neural Tube Defects (Table 4)

The data show that the neural tube defect rate for the state was 8.30 per 10,000 live births. None of the county rates were significantly different from the state rate.

Gastroschisis and Omphalocele (Table 5 and Table 6)

Abdominal wall defects include omphalocele and gastroschisis (Table 5 and Table 6). Young maternal age has been shown to be a high risk factor for gastroschisis.[35] Other risk factors for gastroschisis are maternal use of cocaine, aspirin, or amphetamines; exposure to solvents; and maternal dietary inadequacy.[36,37] Table 5 presents the gastroschisis incidence rate for the state at 3.91 per 10,000 live births. Apache County had the lowest incidence rate, at 0.89, followed by Navajo County, with the rate of 1.48 per 10,000 live births. No other counties had gastroschisis rates significantly different from the state rate. Table 6 presents the omphalocele incidence rate for the state at 2.09 per 10,000 live births. Yuma County had the lowest incidence rate, at 1.01 per 10,000 live births. No county rates were significantly different from the state rate.

Heart Defects (Table 7)

The heart defects included in Table 7 include truncus arteriosus, transposition of the great arteries, tetralogy of Fallot, ventricular septal defect, atrial septal defect, endocardial cushion defect, pulmonary valve atresia and stenosis, tricuspid valve atresia and stenosis, Ebstein's anomaly, aortic valve stenosis, hypoplastic left heart, and coarctation of aorta. The heart defect rate for Gila County was significantly higher than the state rate for heart defects (102.90 vs. 64.86 per 10,000 live births). Navajo County had the second highest heart defect rate, at 75.99, followed by Pima County, at 69.07 per 10,000 live births. The rates for Navajo and Pima Counties were significantly higher than the state rate. On the other hand, Mohave County's prevalence was significantly lower than the state rate, at 51.00 per 10,000 live births.

Table 4
Neural Tube Defects* - Incidence Rates by County, Arizona, 1986-2000
(Live Born Cases Per 10,000 Live Births)

COUNTY	CASES 1986-2000	RATE	95% CONFIDENCE INTERVAL
Arizona	885	8.30	7.75 – 8.85
Apache	18	8.02	4.32-11.73
Cochise	18	7.17	3.86-10.48
Coconino	16	5.73	2.92-8.53
Gila	12	11.99	5.21-18.77
Graham	10	14.96	5.69-24.24
Greenlee	0	0	0
La Paz	4	14.86	0.30-29.42
Maricopa	575	8.92	8.19-9.65
Mohave	17	7.29	3.82-10.75
Navajo	34	12.54	8.33-16.76
Pima	123	8.29	6.92-9.66
Pinal	17	6.82	3.97-9.67
Santa Cruz	5	10.75	4.67-16.84
Yavapai	17	8.09	4.25-11.94
Yuma	19	9.35	6.34-12.36

*Neural tube defects include anencephaly, encephalocele, and spina bifida (see Appendices A and B for definitions).

Table 5
Gastroschisis* - Incidence Rates by County, Arizona, 1986-2000
(Live Born Cases Per 10,000 Live Births)

COUNTY	CASES 1986-2000	RATE	95% CONFIDENCE INTERVAL
Arizona	417	3.91	3.53 – 4.29
Apache	2	0.89	-0.34 – 2.13
Cochise	9	3.58	1.24 – 5.92
Coconino	9	3.22	1.12 – 5.33
Gila	3	3.00	-0.39 – 6.39
Graham	3	4.49	-0.59 – 9.57
Greenlee	2	9.82	-3.79 – 23.44
La Paz	0	0	0 – 0
Maricopa	244	3.97	3.31 – 4.26
Mohave	17	7.29	3.82 – 10.75
Navajo	4	1.48	0.03 – 2.92
Pima	82	4.82	3.78 – 5.86
Pinal	12	3.72	1.61 – 5.82
Santa Cruz	3	2.69	-0.35 – 5.73
Yavapai	12	5.71	2.48 – 8.95
Yuma	15	3.79	1.87 – 5.71

*See Appendices A and B for a definition of gastroschisis and codes included in this diagnostic category.

Table 6
Omphalocele* - Incidence Rates by County, 1986-2000
(Live Born Cases Per 10,000 Live Births)

COUNTY	CASES 1986-2000	RATE	95% CONFIDENCE INTERVAL
Arizona	223	2.09	1.82-2.36
Apache	5	2.23	0.28 – 4.18
Cochise	7	2.79	0.72 – 4.85
Coconino	8	2.86	0.88 – 4.85
Gila	5	5.00	0.62 – 9.37
Graham	0	0	0 – 0
Greenlee	1	4.91	-4.72 – 14.54
La Paz	1	3.71	-3.57 – 11.00
Maricopa	126	1.96	1.61 – 2.30
Mohave	7	3.0	0.78 – 5.22
Navajo	6	2.21	0.44 – 3.98
Pima	41	2.41	1.67 – 3.15
Pinal	5	1.55	0.19 – 2.91
Santa Cruz	2	1.79	-0.69 – 4.28
Yavapai	5	2.38	0.29 – 4.47
Yuma	4	1.01	0.02 – 2.00

*See Appendices A and B for a definition of omphalocele and codes included in this diagnostic category.

Table 7
Heart Defects*- Incidence Rates by County, 1986-2000
(Live Born Cases Per 10,000 Live Births)

COUNTY	CASES 1986-2000	RATE	95% CONFIDENCE INTERVAL
Arizona	6914	64.86	63.33 – 66.39
Apache	137	61.07	50.85-71.30
Cochise	147	58.52	49.06-67.98
Coconino	187	66.94	57.34-76.53
Gila	103	102.90	83.03-122.77
Graham	53	79.31	57.95-100.66
Greenlee	6	29.47	5.89-53.05
La Paz	8	29.72	9.12-50.31
Maricopa	4113	63.83	61.88-65.78
Mohave	119	51.00	41.84-60.16
Navajo	206	75.99	65.61-86.36
Pima	1175	69.07	65.12-73.02
Pinal	200	61.98	53.39-70.58
Santa Cruz	75	67.21	52.00-82.42
Yavapai	149	70.94	59.55-82.33
Yuma	236	59.63	52.02-67.24

*Heart defects include truncus arteriosus, transposition of the great arteries, tetralogy of Fallot, ventricular septal defect, atrial septal defect, endocardial cushion defect, pulmonary valve atresia and stenosis, tricuspid valve atresia and stenosis, Ebstein's anomaly, aortic valve stenosis, hypoplastic left heart, and coarctation of aorta. Appendix B lists codes defining each condition.

APPENDIX A
Definitions of Reported Birth Defects
(As defined in the National Birth Defects Prevention Network's
***Guidelines for Conducting Birth Defect Surveillance*, Appendix 3.2)**

Amniotic Bands
Strands of tissue that float in the amniotic fluid as a consequence of tears or ruptures in the amniotic membrane which surrounds the fetus during development.

Anencephalus
Partial or complete absence of the brain and skull.

Aniridia
Hypoplasia of the iris of both eyes.

Anophthalmia/Microphthalmia
Anophthalmia – Total absence of eye tissue or apparent absence of the globe in an otherwise normal orbit.
Microphthalmia – Reduced volume of the eye. The corneal diameter is usually less than 10 millimeters, or the anteroposterior globe diameter is less than 20 millimeters.

Anotia/Microtia
Anotia – Total absence of the external ear and canal.
Microtia – Malformation or hypoplasia of the external ear (auricle, pinna).

Aortic Valve Stenosis
Obstruction or narrowing of the aortic valve, which may impair blood flow from the left ventricle to the aorta.

Atrial Septal Defect (ASD)
An opening in the septum that separates the left and right atria of the heart.

Biliary Atresia
Congenital absence of the lumen of the extrahepatic bile ducts.

Bladder Exstrophy
A defect in the lower abdominal wall and anterior wall of the bladder through which the lining of the bladder is exposed to the outside.

Choanal Atresia
Congenital obstruction of the opening of the nasal cavity into the nasopharynx on either side. This prevents communication of the nasal cavity with the pharynx.

Cleft Lip with and without Cleft Palate
A defect in the upper lip resulting from incomplete fusion of the parts of the lip.

Cleft Palate without Cleft Lip
An opening in the roof of the mouth resulting from incomplete fusion of the shelves of the palate. The opening may involve the hard palate only, the soft palate only, or both.

Coarctation of the Aorta
Narrowing of the descending aorta, which may obstruct blood flow from the heart to the rest of the body. The most common site of coarctation occurs distal to the origin of the left subclavian artery in the region of the ductus arteriosus.

Common Truncus (Truncus Arteriosus or TA)
Failure of separation of the aorta and the pulmonary artery, resulting in a single common arterial trunk carrying blood from the heart to both the body and lungs.

Congenital Cataract
An opacity of the lens of the eye that has its origin prenatally.

Congenital Hip Dislocation
Location of the head of the femur (bone of the upper leg) outside its normal location in the cup-shaped cavity formed by the hip bones (acetabulum).

Diaphragmatic Hernia
Incomplete formation of the diaphragm through which a portion of the abdominal contents herniate into the thoracic cavity.

Down Syndrome (Trisomy 21)
The presence of three copies of all or a large part of chromosome 21.

Ebstein's Anomaly
Downward displacement if the tricuspid valve into the right ventricle. The tricuspid valve is usually hypoplastic and regurgitant.

Edwards Syndrome (Trisomy 18)
The presence of three copies of all or a large part of chromosome 18.

Encephalocele
Herniation of brain tissue and/or meninges through a defect in the skull. The hernia sac is usually covered by skin.

Endocardial Cushion Defect
A defect in both the lower portion of the atrial septum and the upper portion of the ventricular septum, producing a large opening (canal) in the central part of the heart. The adjacent parts of the mitral and tricuspid valves may also be abnormal, resulting in a single common atrioventricular valve. In extreme cases, virtually the entire atrial and ventricular septae may be missing.

Esophageal Atresia/ Tracheoesophageal Fistula
Esophageal atresia – A condition in which the esophagus ends in a blind pouch and fails to connect with the stomach.
Tracheoesophageal fistula – An abnormal communication between the esophagus and the trachea. This is almost always associated with some form of esophageal atresia.

Fetus/Newborn Affected by Maternal Alcohol Use (Fetal Alcohol Syndrome/FAS)
A spectrum of abnormalities resulting from exposure to alcohol *in utero*. While the specific abnormalities vary among individuals, the hallmarks include growth deficiency, microcephaly, facial dysmorphisms, and neurodevelopmental abnormalities.

Gastroschisis
A congenital opening or fissure in the anterior abdominal wall lateral to the umbilicus through which the small intestine, part of the large intestine, and occasionally the liver and spleen, may herniate. The opening is separated from the umbilicus by a small bridge of skin, and the herniating organs are not covered by a protective membrane. Gastroschisis usually occurs on the right side of the umbilicus, although it may occur on the left.

Hirschsprung Disease (Congenital Megacolon)
Hirschsprung disease – Absence of the parasympathetic ganglion nerve cells (aganglionosis) of the wall of the colon or rectum, which may result in congenital megacolon.
Megacolon – Enlargement of the diameter of part or all of the colon.

Hydrocephalus without Spina Bifida
An increase in the amount of cerebrospinal fluid (CSF) within the brain resulting in enlargement of the cerebral ventricles and increased intracranial pressure.

Hypoplastic Left Heart Syndrome (HLHS)
A condition in which the structures on the left side of the heart and the aorta are extremely small. Classically, this condition includes hypoplasia of the left ventricle, atresia or severe hypoplasia of the mitral and aortic valves, and hypoplasia and coarctation of the aorta.

Hypospadias and Epispadias
Hypospadias – Displacement of the opening of the urethra (urethral meatus) ventrally and proximally (underneath and closer to the body) in relation to the tip of the glans of the penis.
Epispadias – Displacement of the opening of the urethra (urethral meatus) dorsally and proximally (on the top and closer to the body) in relation to the tip of the glans of the penis.

Microcephaly
A cranial vault that is smaller than normal for age. The size of the cranial vault is an indicator of the size of the underlying brain.

Obstructive Genitourinary Defect
Partial or complete obstruction of the flow of urine at any level of the genitourinary tract from the kidney to the urethra.

Omphalocele
A defect in the anterior abdominal wall in which the umbilical ring is widened, allowing herniation of abdominal organs, including the small intestine, part of the large intestine, and occasionally the liver and spleen, into the umbilical cord. The herniating organs are covered by a nearly transparent membranous sac.

Patau Syndrome (Trisomy 13)
The presence of three copies of all or a large part of chromosome 13.

Pulmonary Valve Atresia and Stenosis
Pulmonary valve atresia – Lack of patency, or failure of formation altogether, of the pulmonary valve, resulting in obstruction of blood flow from the right ventricle to the pulmonary artery.
Pulmonary valve stenosis – Obstruction or narrowing of the pulmonary valve, which may impair blood flow from the right ventricle to the pulmonary artery.

Pyloric Stenosis
Hypertrophy (thickening) of the muscles of the pylorus connecting the stomach to the duodenum, resulting in complete or partial obstruction of the passage of food and gastric contents.

Rectal and Large Intestinal Atresia/ Stenosis
Complete or partial occlusion of the lumen of one or more segments of the large intestine and/or rectum.

Reduction Deformity, Lower Limbs
Complete or partial absence of the upper leg (femur), lower leg (tibia and/or fibula), ankle (tarsals), foot (metatarsals), or toes (phalanges).

Reduction Deformity, Upper Limbs
Complete or partial absence of the upper arm (humerus), lower arm (radius and/or ulna), wrist (carpals), hand (metacarpals), or fingers (phalanges).

Renal Agenesis/Hypoplasia
Renal agenesis – Complete absence of the kidney.
Renal hypoplasia – Incomplete development of the kidney.

Spina Bifida without Anencephalus
Incomplete closure of the vertebral spine (usually posteriorly) through which spinal cord tissue and/or the membranes covering the spine (meninges) herniate.

Tetralogy of Fallot
The simultaneous presence of a ventricular septal defect (VSD), pulmonic stenosis, a malpositioned aorta that overrides the ventricular septum, and right ventricular hypertrophy.

Transposition of the Great Arteries (TGA)
Transposition of the aorta and the pulmonary artery such that the aorta arises from the right ventricle (instead of the left) and the pulmonary artery arises from the left ventricle (instead of the right).

Tricuspid Valve Atresia and Stenosis
Tricuspid valve atresia – Lack of patency, or failure of formation altogether, of the tricuspid valve, resulting in obstruction of blood flow from the right atrium to the right ventricle.
Tricuspid valve stenosis – Obstruction or narrowing of the tricuspid valve, which may impair blood flow from the right atrium to the right ventricle.

Ventricular Septal Defect (VSD)
An opening in the septum that separates the left and right ventricles of the heart.

APPENDIX B
ICD-9 and CDC/BPA Codes Defining Conditions in the ABDMP Annual Report

The birth defect categories analyzed in this report are listed below, along with the code ranges that define each category. The center column shows the World Health Organization's International Classification of Disease, 9th Revision, Clinical Modification (ICD-9-CM) (1979) code ranges for each category. The right column specifies the British Pediatric Association Classification of Diseases (BPA) (1979) code ranges for the same conditions.

Condition	ICD-9 codes	BPA codes
Anencephalus	740.0 – 740.1	740.00 – 740.10
Amniotic bands	No code	658.8
Aniridia	743.45	743.42
Anophthalmia/microphthalmia	743.0, 743.1	743.00 – 743.10
Anotia/microtia	744.01, 744.23	744.01, 744.21
Aortic valve stenosis	746.3	746.3
Atrial septal defect	745.5	745.51 – 745.59 (except 745.50)
Biliary atresia	751.61	751.65
Bladder exstrophy	753.5	753.5
Choanal atresia	748	748
Cleft lip with and without cleft palate	749.1, 749.2	749.10 – 749.29
Cleft palate without cleft lip	749	749.00 – 749.09
Coarctation of aorta	747.1	747.10 – 747.19
Common truncus	745	745.00 – 745.01
Congenital cataract	743.30 – 743.34	743.32 – 743.326
Congenital hip dislocation	754.30, 754.31, 754.35	754.3
Diaphragmatic hernia	756.6	756.610 – 756.617
Down syndrome (Trisomy 21)	758	758.00 – 758.09
Ebstein's anomaly	746.2	746.2
Edwards syndrome (Trisomy 18)	758.2	758.20 – 758.290
Encephalocele	742	742.00 – 742.09
Endocardial cushion defect	745.60, 745.61, 745.69	745.60 – 745.69
Esophageal atresia/tracheoesophageal fistula	750.3	750.30 – 750.35
Fetus/newborn affected by maternal alcohol use	760.71	760.71
Gastroschisis	756.79	756.71
Hirshsprung disease	751.3	751.30 – 751.34
Hydrocephalus without spina bifida	742.3 (except 741.0, 741.9)	742.30 – 742.39 (except 741.00 – 741.99)
Hypoplastic left heart syndrome	746.7	746.7
Hypospadias and Epispadias	752.61, 752.62	752.600 – 752.627 (except 752.621)
Microcephalus	742.1	742.1
Obstructive genitourinary defect	753.2, 753.6	753.20-29 – 753.60-69
Omphalocele	756.79	756.7
Patau syndrome (Trisomy 13)	758.1	758.10 – 758.19
Pulmonary valve atresia and stenosis	746.01, 746.02	746.00 – 746.01
Pyloric stenosis	750.5	750.51
Rectal/large intestinal atresia/stenosis	751.2	751.20 – 751.24
Reduction deformity, lower limbs	755.30 – 755.39	755.30 – 755.39
Reduction deformity, upper limbs	755.20 – 755.29	755.20 – 755.29
Renal agenesis/hypoplasia	753	753.00 – 753.01
Spina bifida without anencephalus	741.0, 741.9 (except 740.0 - 740.10)	741.00 – 741.99 (except 740.0 – 740.10)
Tetralogy of Fallot	745.2	745.20 – 745.21, 746.84
Transposition of great arteries	745.10, 745.11, 745.12, 745.19	745.10 – 745.19
Tricuspid valve atresia and stenosis	746.1	746.1 (except 746.105)
Ventricular septal defect	745.4	745.40 – 745.490 (except 745.498)

APPENDIX C
Precision of Diagnosis Codes

Often health care professionals qualify a diagnosis, using words to express their level of confidence that the particular diagnosis explains what has been observed when examining, testing, or performing a procedure on a patient. If a professional makes a diagnosis using a qualifying term, the ABDMP assigns that diagnosis a "precision code," based on the table below. Higher code numbers indicate higher "levels of precision." Generally, if a diagnosis is made several times with different levels of precision, the diagnosis is assigned the precision code consistent with the most certain diagnosis.

Precision Code	Qualifying Terms
1	not stated
2	probably not
3	vs, or
4	rule out, equivocal, questionable, r/o, uncertain, concern for, doubtful
5	suggestive of
6	suspected, suspicious
7	possible, may have, could be, felt to be, perhaps, consider, may be, question of, question
8	consistent with, most likely
9	compatible with, like, appears, evidence of, seems, apparent, believe
10	probable, presume
11	(code not currently used)
12	precise diagnosis, characteristic of (even if qualified with "mild," "somewhat," "relatively," or "borderline")

APPENDIX D
Exclusion List
Non-reportable Birth Defect Cases

The following potential cases are not included in the ABDMP Report:

- Duplicate abstracts and/or duplicated anomalies (cases with multiple abstracts; child seen at more than one facility), i.e., duplicate cases are merged and counted once.
- "Possibles" abstracted for review and consideration and subsequently determined to have conditions or defects that were not reportable according to the ABDMP lists of "excludable conditions."
- Babies born to mothers whose residences are out-of-state or out-of-country (i.e., nonresident cases).
- "Negatives," that is, conditions that were ruled-out during case-finding and medical record review.
- "No match" cases. A birth certificate was not on file and the state of birth could not be confirmed as Arizona.
- Cases among aborted fetuses less than 20 weeks gestation and weighing less than 500 grams. These cases were excluded because there was no reliable denominator that could be used to generate a birth defect rate.
- Prenatally diagnosed cases that did not result in a known live birth or stillbirth are not included. The ABDMP is not currently visiting prenatal diagnostic centers to identify cases.
- Defects with a "precision of diagnosis" code 1-7 are excluded. Only those defects diagnosed at the higher levels of precision (8 or above) are included. Refer to Appendix B for list of precision of diagnosis codes.
- Cases only diagnosed outside of the hospital setting are generally not identified and included in the ABDMP. Children's Rehabilitative Services Clinics and pediatric genetics clinics are the only outpatient facilities we ascertain cases from.

APPENDIX E
Race and Ethnicity

The Arizona Birth Defects Monitoring Program does not collect race and ethnicity data directly from hospital or clinic records. However, all case files are matched with a birth or fetal death certificate prior to being included in the dataset. For statistical purposes, the ABDMP defines the race and ethnicity of the child as equivalent to the mother's race and ethnicity as recorded on the child's birth or fetal death certificate. In this report race and ethnicity combinations are classified the same as in other Arizona Department of Health Services publications, as follows:

Classification of Race/Ethnicity Combinations in Arizona Statistical Reports

	Hispanic	**Non-Hispanic**
White	Hispanic	White
Black	Black	Black
Native American	Native American	Native American
Other	Other	Other

- "White" refers to White non-Hispanics.
- "Hispanic" refers to White Hispanics.
- "Black" refers to African Americans, whether or not they are also Hispanic.
- "Native American" includes people of all Native American tribes, Aleuts, and Eskimos, whether or not they are living on reservation land and whether or not they are Hispanic
- "Other" includes persons who are Asian, unclassified, or did not provide a response to race question on the certificate.

NOTE: Arizona Birth Defects Monitoring Program data provided to the Centers for Disease Control and Prevention (CDC) and which may be published in other documents is analyzed using different classifications for race and ethnicity combinations, as follows.

Classification of Race/Ethnicity Combinations in National Statistical Reports Related to Birth Defects

	Hispanic	**Non-Hispanic**
White	Hispanic	White
Black	Hispanic	Black
Native American	Hispanic	Native American
Other	Other	Other

APPENDIX F
References

1 National Birth Defects Prevention Network (NBDPN). *Guidelines for Conducting Birth Defects Surveillance*. Sever, LE, ed. Atlanta, GA: National Birth Defects Prevention Network, Inc., June 2004.

2 MRC Vitamin Study Research Group. Prevention of neural tube defects: results of the Medical Research Council Vitamin Study. *Lancet.* 1991;338:131–7.

3 Czeizel AE, Dudás I. Prevention of the first occurrence of neural-tube defects by periconceptional vitamin supplementation. *N Engl J Med.* 1992;327:1832–5.

4 Werler, MM, Shapiro, S, and Mitchell, AA. Periconceptional folic acid exposure and risk of occurrent neural tube defects. *JAMA*. 1993;269:1257–61.

5 Centers for Disease Control and Prevention. Spina Bifida and Anencephaly Before and After Folic Acid Mandate --- United States, 1995-1996 and 1999-2000. *MMWR.* 2004; 53(17):362-364.

6 Edmonds LD. Birth Defect Surveillance at the State and Local Level. *Teratology.* 1997; 56:5-7.

7 Yoon PW, et al. The National Birth Defects Prevention Study. *Public Health Rep*. 2001; 116 Suppl 1:32-40.

8 National Birth Defects Prevention Network (NBDPN). *Guidelines for Conducting Birth Defects Surveillance*. Sever, LE, ed. Atlanta, GA: National Birth Defects Prevention Network, Inc., June 2004.

9 Lynberg MC, Edmonds LD. Surveillance of Birth Defects. In: Halperin W, Baker EL, eds. *Public Health Surveillance*. New York, NY: Van Nostrand Reinhold; 1992; 157-177.

10 Erickson JD. Introduction: Birth Defects Surveillance in the United States. *Teratology.* 1997; 65:1-4.

11 Edmonds, LD. Birth Defect Surveillance at the State and Local Level. *Teratology.* 1997; 65:5-7.

12 Lynberg MC, Edmonds LD. Surveillance of Birth Defects. In: Halperin W, Baker EL, eds. *Public Health Surveillance*. New York, NY: Van Nostrand Reinhold; 1992; 157-177.

13 Mrela CK. *Arizona Health Status and Vital Statistics 2004.* Arizona Department of Health Services. September 2005; 147.

14 Mathews TJ, MacDorman MF. Infant Mortality Statistics from the 2003 Period Linked Birth/Infant Death Data Set. *National Vital Statistics Reports.* 2006; 54;(16):8.

15 Centers for Disease Control and Prevention. Years of Potential Life Lost Before Ages 65 and 85 – United States, 1989-1990. *MMWR*; 1992; 41;(18):313-315.

16 Centers for Disease Control and Prevention. Economic costs of Birth Defects and Cerebral Palsy—United States. *MMWR.* 1992; 44;(37):694-699.

17 Graham, JM and Shaw, GM. Gene-Environoment Interactions in Rare Diseases that Include Common Birth Defects. *Birth Defects Research (Part A): Clinical and Molecular Teratology.* 2005; 73:865-867.

18 Khoury, MJ. Genetic Susceptibility to Birth Defects in Humans: From Gene Discovery to Public Health Action. *Teratology*. 2000; 61;17-20.

19 The National Birth Defects Prevention Study: Centers for Birth Defects Research and Prevention Study Protocol. http://www.cdc.gov/ncbddd/protocol /NBDPS_Task_Protocol.pdf (March 23, 2007).

20 Houk VM, Oakley Jr GP, Erickson JD, Mulinare J, and James LM. Recommendations for the Use of Folic Acid to Reduce the Number of Cases of Spina Bifida and Other Neural Tube Defects. *MMWR.* 1992; 41; (RR-14):1-7.

21 Itikala PR, Watkins ML, Mulinare J, Moore CA, Liu Y. Maternal Multivitamin Use and Orofacial Clefts in Offspring. *Teratology.* 2001; 63 (2):79-86.

22 Botto LD, Mulinare J, Erickson JD. Occurrence of Congenital Heart Defects in Relation to Multivitamin Use. *American. Journal of. Epidemiology.* 2000; 151;(9):878-884.

23 Li DK, Daling JR, Mueller BA, Hickok DE, Fantel AG, Weiss NW. Periconceptional Multivitamin Use in Relation to the Risk of Congenital Urinary Tract Anomalies. *Epidemiology.* 1995; 6:212-8.

24 Correa-Villasenor A, Cragan J, Kucik J, O'Leary L, Siffel C, Williams L. "The Metropolitan Atlanta Congenital Defects Program: 35 Years of Birth Defects Surveillance at the Centers for Disease Control and Prevention." *Birth Defects Research (Part A): Clinical and Molecular Teratology*. 2003; 67:617-624.

25 Wasserman CR, Shaw GM, Selvin S, Gould JB, and Syme SL. Socioeconomic Status, Neighborhood Social Conditions, Neural Tube Defects. *American Journal of Public Health.* 1998; 88; (11):1674-1680.

26 Todoroff K, and Shaw GM. Prior Spontaneous Abortion, Prior Elective Termination, Interpregnancy Interval and Risk of Neural Tube Defects. *American Journal of Epidemiology.* 2000 151; (5):505-511.

27 Watkins ML. Efficacy of Folic Acid Prophylaxis for the Prevention of Neural Tube Defects. *Mental Retardation and Developmental Disabilities Research Reviews*. 1998; 4:282-290.

28 Watkins, ML. Efficacy of Folic Acid Prophylaxis for the Prevention of Neural Tube Defects. *Mental Retardation and Developmental Disabilities Research Reviews*. 1998; 4:282-290.

29 Berry RJ, Li Z, Erickson JD, Li S, Moore CA, Wang H, Mulinare J, Zhao P, Wong LY, Gindler J, Hong SX, Correa A. Prevention of Neural Tube Defects with Folic Acid in China (China-U.S. Collaborative Project for Neural Tube Defects), New England Journal of Medicine 1999;341:1485-90.

30 U.S. Department of Health and Human Services. Folic Acid to Fortify U.S. Food Products to Prevent Birth Defects. *HHS News*. February 29, 1996.

31 Centers for Disease Control and Prevention. Spina Bifida and Anencephaly Before and After Folic Acid Mandate --- United States, 1995-1996 and 1999-2000. *MMWR.* 2004; 53(17):362-364.

32 Spina Bifida Association, the National Council of La Raza, GRUMA and Wal-Mart Announce Unprecedented Effort to Fight Spina Bifida. http://www.walmartfacts.com/articles/3828.aspx. Los Angeles, July 11, 2006.

33 Farley TF, Hambridge SJ, and Daley MF. Association of Low Maternal Education with Neural Tube Defects in Colorado, 1989-1998. *Public Health*. 2002; 116:89-94.

34 Shaw GM, Velie EM, and Wasserman CR. Risk for Neural Tube Defect-Affected Pregnancies Among Women of Mexican Descent and White Women of California. *American Journal Public Health.* 1997; 87(9):1467-71.

35 Williams LJ. Kucik JE. Alverson CJ. Olney RS. Correa A. Epidemiology of gastroschisis in Metropolitan Atlanta, 1968 through 2000. *Birth Defects Research. 2005 Mar. 73(3):177-83,* 26

36 Torfs CP, Katz EA, Bateson TF, Lam PK, and Curry CJ. Maternal Medications and Environmental Exposures as Risk Factors for Gastroschisis. *Teratology.* 1996; 54:84-92.

37 Torfs CP, Lam PK, Schaffer DM, and Brand RJ. Association Between Mothers' Nutrient Intake and Their Offspring's Risk of Gastroschisis. *Teratology.* 1998; 58:241-50.