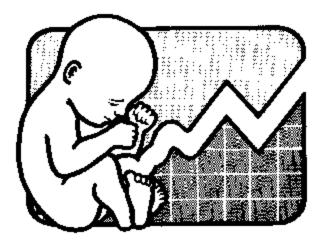


1995 Arizona Birth Defects Monitoring Program Report





Jane Dee Hull Governor State of Arizona

Catherine R. Eden Director Arizona Department of Health Services

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 Epidemiology and Statistics, Bureau of Public Health Statistics at (602) 542-7333.

Permission to quote from or reproduce from this publication is granted when due acknowledgment is made.

Suggested Citation:

Blair, ML. 1995 Arizona Birth Defects Monitoring Program Report. Arizona Department of Health Services, Phoenix, 2000. (Produced as a cooperative effort within the Arizona Birth Defects Monitoring Program.)



1995 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

2700 North Third Street, Ste. #4075 Phoenix, Arizona 85004 (602) 542-7349

June 12, 2000

by

Marilou C. Legazpi Blair, Ph.D. Epidemiologist Office of Epidemiology and Statistics Bureau of Public Health Statistics

ACKNOWLEDGMENTS

Hoa Lien Tran, M.D., M.P.H.Program Manager Matilda ArmentaAbstractor Laura CookAbstractor Kim NguyenAbstractor Ann SniderAbstractor Paula Flores.....Secretary Georgia Armenta Yee, B.S.W., C.T.R....Chief, Office of Health Registries Ross Brechner, M.D., M.S., M.P.H.....Chief, Office of Epidemiology and Statistics Richard Porter, M.S.....Chief, Bureau of Public Health Registries

Tim Flood, M.D.Medical Director

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 accomplish our case finding and abstracting. We also would like to thank Ryan S. Arp and Paul A. Barbeau, GIS consultants for developing the maps for this report. The Arizona Birth Defects Monitoring Program welcomes comments, questions, and observations.

TABLE OF CONTENTS

Page

| Executive Summary |
|--|
| The Importance of Arizona's Birth Defects Registry |
| Methods |
| Case Definition |
| Interpreting the Data |
| State Profile of Defects |
| Race/Ethnicity |
| Maternal Age |
| County Profiles |
| Sentinel Defects |

List of Tables

| Table 1. Congenital Anomalies by Race/Ethnicity - Arizona 1995 10 |
|--|
| Table 2. Birth Defects Totals by County of Residence, 1995 12 |
| Table 3. Congenital Anomalies by Year, 1986 - 1992, 19951995 |
| Table 4. Birth Defect Rates by County, 1986 - 1992, 1995 38 |
| Table 5. Birth Defects Rates by Race/Ethnicity by County, 1986 - 1992, 1995 39 |
| Table 6. Chromosomal Defects - Rates by County, 1986 - 1992,1995 |
| Table 7. Oral Clefts - Rates by County, 1986 - 1992, 1995 |
| Table 8. Neural Tube Defects - Rates by County, 1986 - 1992, 1995 |
| Table 9. Abdominal Wall Defects - Rates by County, 1986 - 1992, 1995199547 |
| Table 10. Heart Defects - Rates by County, 1986 - 1992, 1995 1995 48 |

TABLE OF CONTENTSList of Figures

| Figure 1. Leading Causes of Infant Mortality in the U.S., 1998 |
|---|
| Figure 2. Trends for Congenital Anomalies, 1986 - 1992, 1995 |
| Figure 3. Spina Bifida Incidence Rates by Race/Ethnicity, 1995 |
| Figure 4. Abdominal Wall Defect Incidence Rates by Race/Ethnicity, 1995 |
| Figure 5. Down Syndrome Incidence Rates by Race/Ethnicity, 1995 |
| Figure 6. Microcephaly Incidence Rates by Race/Ethnicity, 1995 |
| Figure 7. Pyloric Stenosis Incidence Rates by Race/Ethnicity, 1995 |
| Figure 8. Birth Defects Rates by Maternal Age |
| Figure 9. Down Syndrome (Trisomy 21) by Maternal Age |
| Figure 10. Gastroschisis by Maternal Age |
| Figure 11. Oral Clefts Incidence Rates by County, Arizona, 1986-1992 & 1995 |
| Figure 12. Oral Clefts Cases, Arizona, 1995 |

Appendices

| Appendix 1. Conditions Included in Figures |
|--|
| Appendix 2. Conditions Shown in Tables |
| Appendix 3. Precision of Diagnosis codes |
| Appendix 4. Abbreviations |
| Appendix 5. Exclusion list |
| Appendix 6. References |
| Appendix 7. Birth Weight |
| Appendix 8. Other Birth Defects Collected, Arizona, 1995 |

EXECUTIVE SUMMARY

There were 1,386 children with a reportable birth defect born to Arizona residents in Arizona in 1995. During this period there were 72,386 live births and 497 still births in Arizona. This report presents 44 composite categories of birth defects developed by the Centers for Disease Control and Prevention (CDC) (Appendix 2). These categories are the most serious defects. Of the 1,386 children, 1,013 are included in this report for these 44 categories. Arizona's overall birth defect rate is 19.0 cases per 1,000 births in 1995, which is slightly higher than the 1992 birth defect rate of 18.26 cases per 1,000 births and lower than 1991 birth defect rate of 31.4 cases per 1,000 births. It should be noted that the decrease in the birth defect rate between 1991 and 1992 is due to a reduction in the number of reportable birth defects conditions included in the Arizona Birth Defects Monitoring Program (ABDMP) from 500 to 140, effective with 1992 births. The most common birth defects observed were pyloric stenosis (148 cases), oral clefts (141 cases), obstruction of kidney/ureter (108 cases), Down syndrome (90 cases), dislocation of hip (83 cases), and microcephaly (81 cases) (Table 1). Other common defects are hypospadias (179 cases) and ventricular septal defect (141 cases) (Appendix 8).

Race/Ethnicity Patterns

Native Americans had the highest rates for microcephaly. The rates of pyloric stenosis was highest for Hispanics, followed by Blacks and Whites. Spina bifida was the most common neural tube defect (NTD) among all races; however, it was most common among Hispanics. Down syndrome exhibited highest rates among Blacks. Tests of significance indicate that none of these differences are statistically significant.

Age Patterns

Observed rates for all birth defects were highest among women 35 years of age and older. The rate of Down syndrome (Trisomy 21) increased with maternal age. Gastroschisis, an abdominal wall defect, showed highest rates among young mothers and decreased in incidence with maternal age.

County Patterns

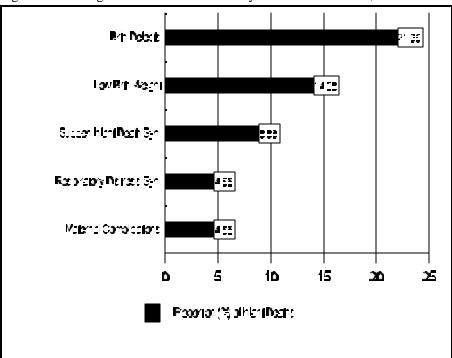
Birth defects data are presented by county. Cases are aggregated for the years 1986 through 1992 and 1995 to provide numbers large enough for analysis. Gila county had the highest rate of congenital anomalies, whereas Greenlee and La Paz counties had the lowest rates. The results of the z-tests indicate that the overall birth defect rate of Gila county is significantly higher than the state rate (p < .01). The 44 composite categories of birth defects were also examined by race and county. Statistically significant differences were found only for Apache county rates for whites and Gila county rates for Native Americans. The results show that Apache county rates for Whites is significantly lower at the 0.01 level than the state rate for whites. In contrast, the Gila county rates for Native Americans is significantly higher than the state rate (p<.01). The rates of five sentinel defects (chromosomal defects, oral clefts, heart defects, abdominal wall defects, and neural tube defects) are examined by county. The county rates are not significantly different from the state rate for any of these defects. Chromosomal defect rates are highest for Gila county (2.28 per 1,000 live births) and Navajo county (1.91 per 1,000 births) and lowest for Greenlee, La Paz, Graham, Santa Cruz, and Mohave counties. For oral clefts, Apache and Gila counties have the highest rates for this time period at 3.05 and 2.84 per 1,000 live births respectively while, Greenlee, La Paz, Graham and Maricopa countries have the lowest rates. Maps are used to show the incidence rates of oral clefts for 1986 to 1992 and 1995 and the distribution of oral cleft cases for 1995. The maps show that oral cleft cases are widely dispersed across the state. Neural tube defects rates is highest for Navajo county at 1.25 per 1,000 live births, but is not statistically different from the state rate. Mohave county has the highest incidence rates for abdominal wall defects at 0.89 per 1,000 live births in contrast to the rate at the state level (0.50 per 1,000 live births), but is not statistically different from the state rate. Heart defect rates are highest for Navajo and Gila counties at 2.11 and 2.09 per 1,000 live births. These rates, however, are statistically different from the state rate of 1.44 per 1,000 live births at (p<.01).

THE IMPORTANCE OF ARIZONA'S BIRTH DEFECTS REGISTRY

The Arizona Birth Defects Monitoring Program (ABDMP) serves several public health functions. It is a population-based registry which provides accurate counts used for prevention efforts, planning health services, and ongoing surveillance to monitor for trends and early detection of problems. Such a registry is necessary because other systems for reporting birth defects, including birth certificates and hospital discharge data are not accurate or complete due to under reporting in the number of cases, lack of specificity of birth defects, and possible incomplete recording of birth defects information.¹ In addition, research shows that birth certificates often indicate defects that in fact were not present. $_{2,3,4,5,6,7}$

Economic Impact

Birth defects are the leading cause of infant mortality in the United States. In 1998, 22.0 percent of infant deaths were due to birth defects (see Figure 1).⁸ In addition, birth defects are the fifth leading cause of years of potential life lost. The most recent study of population-based hospital discharge data in two states show that 12 percent of pediatric admissions were associated with birth defects and genetic diseases in 1991. Total hospital charges for these admissions were 2.84 times higher than





charges for children who were admitted for all other reasons combined.⁹ Another study using California's population based data (adjusted to provide national estimates) and national data estimated the economic cost of the most clinically important structural birth defects in the United States. Their results show that the combined estimated costs of 18 structural defects in the United States in 1992 was \$8 billion, with costs ranging from \$75,000 to \$503,000 per new case. Birth defects which were characterized by relatively high levels of long-term activity limitation had higher costs (i.e. Down Syndrome (\$451,000), the heart defect Truncus Arteriosus, (\$503,000), and Spina Bifida (\$294,000)).¹⁰ Harris and James produced these estimates for each state and included factors such as lost wages to family members caring for children who have birth defects and psychosocial costs. Data on the 1988 Arizona birth cohort show that the estimated lifetime costs in1992 dollars of selected birth defects in Arizona range from \$1,275,543 to \$41,596,118, depending on the defect category.¹¹

Human Cost

While the economic costs associated with birth defects are easier to ascertain, estimating the human and societal costs is more difficult. Human and societal costs of birth defects are usually reflected by the impact of birth defects on infant mortality and the number of years of potential life lost. Case ascertainment systems that use intensive measures to document birth defects find that an estimated 3-5 percent of births have a serious birth defect. This would imply that 116,000 to 194,000 babies in the United States in 1997 were born with a serious birth defect. ¹² For Arizona, in 1995 there was a total of 1,386 live births and fetal deaths with a birth defect.

Scientists know the cause of only a relatively small number of defects. For example, maternal alcohol consumption causes fetal alcohol syndrome; German measles in early pregnancy causes congenital rubella syndrome. There may be many defects caused by teratogens, yet to be discovered. However, the search for causes of birth defects is a difficult process. If Arizona is to ensure its children a healthy future, we must continue to search for the causes of congenital anomalies. Also, birth defect registries are a vital first step in reducing birth defects. The documentation of baseline birth defect rates in Arizona provides the starting point against which we can measure successful interventions.

METHODS

Data Sources and Procedures

The ABDMP is a statewide, population-based, active surveillance program, pursuant to ARS §36-133 which mandates the surveillance of chronic diseases, including birth defects. Trained ABDMP staff collect data from 64 reporting sources: 58 hospitals, including Phoenix Children's Hospital; 2 centers providing genetics services; 4 clinics of the state Children's Rehabilitative Services; and the state Office of Vital Records. Ascertainment procedures used by the ABDMP are nearly identical to those used by the California Birth Defects Monitoring Program and the US Centers for Disease Control's Metropolitan Atlanta Congenital Defects Program (MACDP).

Sources of data at hospitals include the disease index, labor and delivery log, nursery log, newborn intensive care log, pediatric log, pathology/autopsy log. Not all sources are available at each hospital. Potential cases are identified through a review of the hospital's disease index and various logs. This process is called case finding. Next, the medical records of possible cases are pulled and reviewed to determine which records meet the case definition. An abstract of the medical record then is completed for each reportable case. In order to find the birth defect cases born in 1995, ABDMP staff reviewed more than 10,000 medical records, identified reportable cases, and excluded those not meeting the case definition.

In addition to the hospital sources, Certificates of Birth, Death, and Fetal Death that indicate a birth defect are reviewed and matched against cases listed in the registry. Medical records then are requested from the reporting hospitals on those children not previously identified from other sources and if the condition(s) reported meet the case definition, pertinent information is abstracted for the registry. 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 (which occurs 2-4 years after the child's birth or fetal death) then the more precise diagnosis is used.

The abstracts of cases identified from multiple sources are compared, merged, and added to the registry. Inconsistencies, differences and/or conflicting data are resolved before being entered into the ABDMP system.

ABDMP staff assign a six-digit classification code to each defect. The classification system is CDC's modification of the British Pediatric Association (BPA) Classification of Disease. This coding system is similar to the International Classification of Disease (ICD). The staff collect diagnostic information on

birth defects that fall primarily within the range of ICD-9-CM Codes 740.00-759.99. The system of codes is hierarchical: the more digits in the code, the more precise the diagnosis. ABDMP staff always code the data at the most precise level possible.

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 structural, genetic, or biochemical birth defect, or other specified birth outcome that can adversely affect an infant's health and development (most, but not all, are listed in ICD-9-CM 740.0-759.9).
- C. The defect must be diagnosed, or signs and symptoms of a potential defect recognized, within the first year of life.
- D. Stillborn infants are included if they have a reportable birth defect.
- E. The date of birth (or delivery for stillbirths > 19 weeks of gestational age) is on or after January 1, 1986.

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 to include only the major congenital anomalies recommended by "The International Clearinghouse for Birth Defects Monitoring Systems" and recommended by CDC. The reduced list of reportable defects went into effect starting with births occurring in 1992. The retained, reportable defects still permit the ABDMP to compare its rates with other registries for the major birth defects categories. The number of reportable congenital anomalies was reduced from over 500 to 140 conditions.

Operationally, the ABDMP staff collected data for the births occurring in 1992 and 1995 at about the same time. This was done to expedite the registration of these defects.

INTERPRETING THE DATA

The tables and figures presented in this report represent data collected on birth defects in Arizona for the period 1986 to 1992, and 1995. 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 were diagnosed with a particular reportable birth defect within the first year of life. Children born with more than one reportable defect, as often occurs, are listed simultaneously in as many of the 44 selected birth defect categories as are applicable. However, within any one of the 44 categories, a child is listed no more than once.

Rates 1 4 1

Incidence rates of birth defects were calculated by dividing the number of children 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. In most tables and figures, we show rates that are calculated by including live births and fetal deaths in both the numerator and denominator. For example, there were 90 cases (live born and still born infants \geq 20 weeks of gestation) of Down Syndrome in 1995. There were 72,883 births (either live births or fetal deaths) in 1995. The rate is calculated as such: (90/72883)*10,000 = 12.35 cases of Down Syndrome per 10,000 live births and fetal deaths.

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 99 percent Poisson confidence intervals. The confidence intervals indicate that the true rate should be contained in this interval 99 percent of the time. For example, Down Syndrome occurs at a rate of 12.35 per 10,000 births. The lower and upper bounds of the point estimate of this rate are 9.2 and 16.1, respectively. Thus, one can say that 99 percent of the time that the true rate of Down Syndrome is between 9.2 and 16.1 cases per 10,000 live births and fetal deaths.

Small Numbers and a Note Of Caution

While the intent of these data is to provide the reader with useful information on birth defects in Arizona, an equally important point is not to mislead data users. Therefore, it is important to stress that rates, confidence intervals, or any other analysis based on fewer than 10 reported cases cannot be considered statistically reliable.

STATE PROFILE OF DEFECTS

State Data

This is the eighth annual report of data compiled by the ABDMP in its mission to collect and analyze information on children with birth defects and to provide data for the study of causes of birth defects in Arizona.

Tables and Figures

Table 1 presents data on 44 selected congenital anomalies by race for 1995. Table 2 looks at all reportable birth defects for both live births and fetal deaths. Fetal deaths include therapeutic abortions and still-born babies with a reportable congenital defect if the estimated gestational age is greater than 19 weeks. Table 3 displays birth defect rates by year for 1986 through 1992 and 1995. The series of graphs in Figure 2 display the trends for selected congenital anomalies.

County and Race/Ethnicity

An expanded look at selected birth defects and race/ethnicity follow the section on state profile. County level data is presented later in this report under the heading County Profiles.

| Table 1 |
|--|
| Arizona Birth Defects Monitoring Program |
| Congenital Anomalies - Arizona 1995 |
| Incidence Rates ^{a,b} per 10,000 Live Births and Fetal Deaths |

| CODE | DEFECT GROUP | <u>TOTAL</u> | RATE | <u>WHITE</u> | RATE | HISP. | RATE | <u>BLACK</u> | RATE | NATIVE AMER. | RATE | <u>OTHER</u> | RATE |
|--|---|--------------------------------------|--|----------------|--|------------------------------------|--|------------------|---|-----------------------|--|-----------------------|--|
| | CENTRAL NERVOUS SYSTEM Anencephaly Spina Bifida w/ Hydrocephaly Spina Bifida w/o Hydrocephaly Encephalocele Hydrocephaly Microcephaly | 18 24 10 7 40 81 | 2.47 3.29 1.37 0.96 5.49 11.11 | 4 1 | 1.55 2.58 1.03 0.26 4.13 6.20 | 8 11 4 4 18 38 | 3.16 4.35 1.58 1.58 7.11 15.02 | 2 1 0 | 0.00 8.83 4.41 0.00 0.00 26.48 | 1 1 2 5 | 3.89 1.95 1.95 3.89 9.72 23.34 | 0 0 0 1 | 0.00 0.00 0.00 0.00 6.92 6.92 |
| B00 B03 B04 B51 B52 B54 | EYE AND EAR Glaucoma Cataract Anophthalmia Microphthalmia Ear Anomaly w/ hearing loss | 5 14 2 24 46 | 0.69 1.92 0.27 3.29 6.31 | 9 2 11 | 1.03 2.32 0.52 2.84 4.65 | 1 2 0 7 21 | 0.39 0.79 0.00 2.77 8.30 | 2 0 | 0.00 8.83 0.00 0.00 4.41 | 1 0 | 0.00 1.95 0.00 11.67 11.67 | 0 0 0 | 0.00 0.00 0.00 0.00 0.00 |
| D02 D03 D04 D51 D52 | CARDIAC Truncus Arteriosus Transposition of great vessels Tetralogy of Fallot Single ventricle Aortic stenosis Hypoplastic left heart Tot. anomal. pulm. ven. return | 3 33 29 5 30 10 12 | 0.41 4.53 3.98 0.69 4.12 1.37 1.65 | 3 17 7 | 0.77 3.61 2.58 0.77 4.39 1.81 0.52 | 0 13 13 1 10 3 7 | 0.00 5.14 5.14 0.40 3.95 1.18 2.77 | 3 2 0 0 | 0.00 13.24 8.83 0.00 0.00 0.00 0.00 | 2 3 1 3 0 | 0.00 3.89 5.83 1.95 5.83 0.00 5.83 | 1 1 0 0 0 | 0.00 6.92 6.92 0.00 0.00 0.00 0.00 |
| E00 E01 E06 | RESPIRATORY Choanal atresia Agenesis of lung | 14 14 | 1.92 1.92 | 8 8 | 2.06 2.06 | | 1.58 1.58 | | 4.41 8.83 | 1 0 | 1.94 0.00 | - | 0.00 |
| F01 F08 F09 | OROFACIAL AND GASTROINTESTINAL Cleft palate Cleft lip w&wo cleft palate Pyloric stenosis Tracheo-esophageal fistula | 47 94 148 18 | 6.45 12.90 20.31 2.47 | 46 75 13 | 6.71 11.88 19.36 3.36 | 5 | 6.32 13.04 24.51 3.36 | 4 5 0 | 17.65 17.65 22.06 0.00 | 11 4 0 | 1.94 21.39 7.78 0.00 | 0 2 0 | 0.00 0.00 13.83 0.00 |

^a Incidence rates include live-born and still born cases. ^b Incidence rates based on counts of less than 10 events are not statistically reliable.

Table 1 Continued Arizona Birth Defects Monitoring Program Congenital Anomalies - Arizona 1995 Incidence Rates^{a,b} per 10,000 Live Births and Fetal Deaths

| CODE | DEFECT GROUP | TOTAL | <u>RATE</u> | <u>WHITE</u> | <u>RATE</u> | <u>HISP.</u> | RATE | <u>BLACK</u> | <u>RATE</u> | NATIVE AMER. | <u>RATE</u> | <u>OTHER</u> | <u>RATE</u> |
|--|---|--------------------------------------|---|------------------------|--|-----------------------------|---|----------------------------|--|-----------------------|---|--------------|---|
| F00 F14 F15 F16 F17 F18 F21 | OROFACIAL AND GASTROINTESTINAL Stenosis/atresia of duodenum Stenosis/atresia of sm. intest Stenosis/atresia of rectum Hirschsprung's disease Malrotation of intestine Biliary atresia | 9 11 37 16 19 3 | 1.23 1.51 5.08 2.19 2.61 0.41 | 4 17 | 0.52 1.03 4.39 1.29 3.10 0.26 | 6 5 15 9 6 1 | 2.37 1.98 5.94 3.56 2.37 0.39 | 0 2 1 1 1 | 0.00 8.83 8.83 4.41 4.41 4.41 | 0 3 | 1.94 0.00 5.83 0.00 0.00 0.00 | 0 | 0.00 0.00 6.92 0.00 0.00 |
| H00 H01 H06 H09 | GENITO-URINARY Renal agenesis Obstruction of kidney/ureter Bladder or urethra obstruction | 39 108 7 | 5.35 14.82 0.96 | 45 | 5.42 11.62 0.52 | 16 48 5 | 6.32 18.97 1.98 | 1 1 0 | 4.41 4.41 0.00 | 10 | 1.94 19.45 0.00 | | 0.00 27.66 0.00 |
| J00 J03 J51 J52 K05 N01 N02 N04 | MUSCULOSKELETAL Dislocation of hip Complete absence upp/low limb Phocomelia of Limb Amniotic Bands Diaphragmatic hernia Omphalocele Gastroschisis | 83 2 0 12 20 14 27 | 11.39 0.27 0.00 1.65 2.74 1.92 3.70 | 1 0 7 13 7 | 8.26 0.26 0.00 1.81 3.36 1.81 3.61 | 33 0 5 5 4 8 | 13.04 0.00 1.98 1.98 1.58 3.16 | 2 0 0 1 1 2 | 8.83 0.00 0.00 4.41 4.41 8.83 | 1 0 0 1 2 | 27.23 1.94 0.00 0.00 1.94 3.89 5.83 | 0 | 13.83 0.00 0.00 0.00 0.00 0.00 0.00 |
| R00 R01 R02 R03 S02 | SYNDROMES Down Syndrome (Trisomy 21) Patau Syndrome (Trisomy 13) Edwards Syndrome (Trisomy 18) Fetal Alcohol Syndrome | 90 8 18 27 | 12.35 1.10 2.47 3.70 | 4 9 | 11.62 1.03 2.32 1.03 | 34 4 5 | 13.44 1.58 2.37 1.98 | 5 0 1 2 | 22.06 0.00 4.41 8.83 | 0 2 | 7.78 0.00 3.89 29.17 | 0 0 | 13.83 0.00 0.00 6.92 |

^a Incidence rates include live born and still born cases. ^b Incidence rates based on counts of less than 10 events are not statistically reliable.

Table 2

Arizona Birth Defects Monitoring Program ^{a, b} Birth Defects by County of Residence, 1995 (140 Conditions Monitored)

| STATE, COUNTY | - | BIRTHS FECTS | STILL W/ DE | BIRTHS FECTS | - | D STILL FECTS | DEFEC | NUMBER OF DEFECTS OF LIVE BIRTHS | | ER OF TS OF BIRTHS | |
|-------------------|--------|-----------------|----------------|-----------------|--------|------------------|--------|--|--------|--------------------------|--|
| | Number | % OF LB | Number | % OF SB | Number | % TOT. | Number | AVG Number | Number | AVG Number | |
| ARIZONA | 1330 | 1.84 | 56 | 11.27 | 1386 | 1.90 | 2279 | 1.71 | 119 | 2.12 | |
| APACHE COUNTY | 24 | 1.80 | 0 | 0.00 | 24 | 1.79 | 38 | 1.58 | 0 | 0.00 | |
| COCHISE COUNTY | 27 | 1.54 | 0 | 0.00 | 27 | 1.53 | 45 | 1.67 | 0 | 0.00 | |
| COCONINO COUNTY | 32 | 1.82 | 1 | 6.67 | 33 | 1.86 | 59 | 1.84 | 1 | 1.00 | |
| GILA COUNTY | 15 | 2.19 | 1 | 25.00 | 16 | 2.32 | 29 | 1.93 | 1 | 1.00 | |
| GRAHAM COUNTY | 7 | 1.76 | 0 | 0.00 | 7 | 1.76 | 10 | 1.43 | 0 | 0.00 | |
| GREENLEE COUNTY | 3 | 1.91 | 0 | 0.00 | 3 | 1.91 | 10 | 3.33 | 0 | 0.00 | |
| LA PAZ COUNTY | 4 | 2.09 | 1 | 50.00 | 5 | 2.59 | 4 | 1.00 | 9 | 9.00 | |
| MARICOPA COUNTY | 825 | 1.87 | 42 | 13.64 | 867 | 1.96 | 1373 | 1.66 | 83 | 1.98 | |
| MOHAVE COUNTY | 32 | 1.74 | 2 | 16.67 | 34 | 1.83 | 53 | 1.66 | 5 | 2.50 | |
| NAVAJO COUNTY | 44 | 2.66 | 1 | 6.25 | 45 | 2.70 | 88 | 2.00 | 2 | 2.00 | |
| PIMA COUNTY | 179 | 1.60 | 8 | 10.96 | 187 | 1.66 | 345 | 1.93 | 18 | 2.25 | |
| PINAL COUNTY | 37 | 1.82 | 0 | 0.00 | 37 | 1.81 | 60 | 1.62 | 0 | 0.00 | |
| SANTA CRUZ COUNTY | 16 | 2.03 | 0 | 0.00 | 16 | 2.02 | 30 | 1.87 | 0 | 0.00 | |
| YAVAPAI COUNTY | 24 | 1.57 | 0 | 0.00 | 24 | 1.56 | 51 | 2.12 | 0 | 0.00 | |
| YUMA COUNTY | 61 | 2.00 | 0 | 0.00 | 61 | 1.99 | 84 | 1.38 | 0 | 0.00 | |

^aTotal number of live births in Arizona for 1995 = 72,386

^bTotal number of fetal deaths in Arizona for 1995 = 497

Table 3 Arizona Birth Defects Monitoring Program Incidence Rates^a Per 1,000 Live Births and Fetal Deaths, Arizona

| CODE/CONDITION (1) | | | | | | | | | |
|-------------------------------|-------|-------------|-------------|-------------|-------------|-------------|-------------|-------------|-------------|
| | | <u>1986</u> | <u>1987</u> | <u>1988</u> | <u>1989</u> | <u>1990</u> | <u>1991</u> | <u>1992</u> | <u>1995</u> |
| A01 Anencephaly | Cases | 22 | 17 | 18 | 18 | 16 | 17 | 21 | 18 |
| | Rate | 0.35 | 0.26 | 0.27 | 0.27 | 0.23 | 0.25 | 0.30 | 0.25 |
| | CI | 0.19-0.60 | 0.12-0.48 | 0.13-0.48 | 0.13-0.48 | 0.11-0.43 | 0.12-0.45 | 0.16-0.52 | 0.12-0.44 |
| A02 S.B. with Hydrocephaly | Cases | 26 | 24 | 19 | 22 | 23 | 21 | 26 | 24 |
| | Rate | 0.42 | 0.37 | 0.28 | 0.33 | 0.33 | 0.31 | 0.38 | 0.33 |
| | CI | 0.24-0.69 | 0.20-0.62 | 0.14-0.50 | 0.17-0.55 | 0.18-0.56 | 0.16-0.53 | 0.21-0.61 | 0.18-0.55 |
| A03 S.B. without Hydrocephaly | Cases | 10 | 11 | 6 | 16 | 16 | 13 | 12 | 10 |
| | Rate | 0.16 | 0.17 | 0.09 | 0.24 | 0.23 | 0.19 | 0.17 | 0.14 |
| | CI | 0.06-0.35 | 0.06-0.35 | 0.02-0.23 | 0.11-0.44 | 0.11-0.43 | 0.08-0.37 | 0.07-0.35 | 0.05-0.29 |
| A13 Encephalocele | Cases | 10 | 8 | 14 | 5 | 13 | 14 | 2 | 7 |
| | Rate | 0.16 | 0.12 | 0.21 | 0.07 | 0.19 | 0.20 | 0.03 | 0.10 |
| | CI | 0.06-0.35 | 0.03-0.29 | 0.09-0.40 | 0.02-0.21 | 0.08-0.37 | 0.09-0.39 | 0.00-0.13 | 0.03-0.24 |
| A15 Hydrocephaly | Cases | 34 | 41 | 48 | 44 | 52 | 46 | 34 | 40 |
| | Rate | 0.55 | 0.64 | 0.72 | 0.65 | 0.75 | 0.67 | 0.49 | 0.55 |
| | CI | 0.34-0.85 | 0.41-0.95 | 0.48-1.04 | 0.43-0.95 | 0.51-1.06 | 0.44-0.97 | 0.30-0.75 | 0.35-0.82 |
| A16 Microcephaly | Cases | 30 | 60 | 70 | 109 | 118 | 120 | 90 | 81 |
| | Rate | 0.49 | 0.94 | 1.06 | 1.61 | 1.70 | 1.75 | 1.30 | 1.11 |
| | CI | 0.29-0.77 | 0.65-1.30 | 0.76-1.43 | 1.17-1.96 | 1.33-2.15 | 1.37-2.21 | 0.97-1.70 | 0.82-1.47 |
| B03 Glaucoma | Cases | 2 | 7 | 4 | 5 | 4 | 2 | 1 | 5 |
| | Rate | 0.03 | 0.10 | 0.06 | 0.07 | 0.06 | 0.03 | 0.01 | 0.07 |
| | CI | 0.04-0.15 | 0.03-0.26 | 0.00-0.19 | 0.02-0.21 | 0.01-0.18 | 0.00-0.14 | 0.00-0.11 | 0.01-0.19 |

| Table 3 Continued |
|--|
| Arizona Birth Defects Monitoring Program |
| Incidence Rates ^a Per 1,000 Live Births and Fetal Deaths, Arizona |

| CODE/CONDITION (1) | | | | | | | | | |
|---------------------------------------|---------------------|-------------------------|-------------------------|-------------------------|-------------------------|-------------------------|-------------------------|-------------------------|-------------------------|
| | | <u>1986</u> | <u>1987</u> | <u>1988</u> | <u>1989</u> | <u>1990</u> | <u>1991</u> | <u>1992</u> | <u>1995</u> |
| B04 Cataract | Cases | 8 | 7 | 7 | 15 | 24 | 10 | 12 | 14 |
| | Rate | 0.13 | 0.10 | 0.10 | 0.22 | 0.35 | 0.15 | 0.17 | 0.19 |
| | CI | 0.04-0.30 | 0.03-0.26 | 0.03-0.26 | 0.10-0.42 | 0.19-0.57 | 0.05-0.31 | 0.07-0.35 | 0.09-0.37 |
| B51 Anophthalmia | Cases | 6 | 1 | 3 | 5 | 7 | 5 | 3 | 2 |
| | Rate | 0.09 | 0.01 | 0.04 | 0.07 | 0.10 | 0.07 | 0.04 | 0.03 |
| | CI | 0.02-0.25 | 0.00-0.11 | 0.00-0.16 | 0.02-0.21 | 0.03-0.25 | 0.02-0.21 | 0.00-0.16 | 0.00-0.13 |
| B52 Microphthalmia | Cases | 10 | 24 | 21 | 19 | 24 | 29 | 22 | 24 |
| | Rate | 0.16 | 0.37 | 0.31 | 0.28 | 0.35 | 0.42 | 0.32 | 0.33 |
| | CI | 0.06-0.35 | 0.20-0.62 | 0.16-0.54 | 0.14-0.50 | 0.19-0.57 | 0.25-0.67 | 0.17-0.54 | 0.18-0.55 |
| B54 Hearing loss w/ear anomaly | Cases | 33 | 59 | 34 | 50 | 59 | 65 | 41 | 44 |
| | Rate | 0.53 | 0.92 | 0.51 | 0.74 | 0.85 | 0.95 | 0.59 | 0.60 |
| | CI | 0.32-0.83 | 0.64-1.28 | 0.31-0.79 | 0.50-1.06 | 0.59-1.18 | 0.67-1.30 | 0.38-0.88 | 0.39-0.88 |
| D01 Truncus Arteriosus | Cases | 4 | 10 | 9 | 9 | 6 | 6 | 3 | 3 |
| | Rate | 0.06 | 0.15 | 0.13 | 0.13 | 0.09 | 0.09 | 0.04 | 0.04 |
| | CI | 0.01-0.20 | 0.05-0.33 | 0.04-0.30 | 0.05-0.30 | 0.02-0.23 | 0.02-0.23 | 0.00-0.16 | 0.00-0.15 |
| D02 Transposition of Great Vessels | Cases Rate CI | 32 0.52 0.31-0.81 | 26 0.40 0.23-0.66 | 26 0.39 0.22-0.64 | 33 0.49 0.30-0.75 | 28 0.40 0.23-0.65 | 26 0.38 0.21-0.62 | 25 0.36 0.20-0.59 | 33 0.45 0.28-0.70 |
| D03 Tetralogy of Fallot | Cases | 15 | 18 | 29 | 23 | 27 | 22 | 32 | 29 |
| | Rate | 0.24 | 0.28 | 0.43 | 0.34 | 0.39 | 0.32 | 0.46 | 0.40 |
| | CI | 0.11-0.46 | 0.13-0.50 | 0.25-0.69 | 0.19-0.57 | 0.22-0.63 | 0.17-0.54 | 0.28-0.72 | 0.23-0.63 |

Table 3 Continued Arizona Birth Defects Monitoring Program Incidence Rates^a Per 1,000 Live Births and Fetal Deaths, Arizona

| CODE/CONDITION (1) | | | | | | | | | |
|--|---------------------|------------------------|------------------------|-------------------------|-------------------------|-------------------------|-------------------------|-------------------------|-------------------------|
| | | <u>1986</u> | <u>1987</u> | <u>1988</u> | <u>1989</u> | <u>1990</u> | <u>1991</u> | <u>1992</u> | <u>1995</u> |
| D04 Single Ventricle | Cases | 2 | 4 | 5 | 4 | 6 | 1 | 3 | 5 |
| | Rate | 0.03 | 0.06 | 0.07 | 0.06 | 0.09 | 0.01 | 0.04 | 0.07 |
| | CI | 0.00-0.15 | 0.01-0.19 | 0.01-0.21 | 0.01-0.19 | 0.02-0.23 | 0.00-0.11 | 0.00-0.16 | 0.01-0.19 |
| D51 Aortic Stenosis | Cases | 8 | 15 | 17 | 25 | 17 | 17 | 23 | 30 |
| | Rate | 0.13 | 0.23 | 0.25 | 0.37 | 0.25 | 0.25 | 0.32 | 0.41 |
| | CI | 0.04-0.30 | 0.10-0.44 | 0.12-0.46 | 0.21-0.61 | 0.12-0.45 | 0.12-0.45 | 0.18-0.56 | 0.24-0.65 |
| D52 Hypoplastic Left Heart | Cases | 9 | 16 | 8 | 16 | 19 | 11 | 13 | 10 |
| | Rate | 0.14 | 0.25 | 0.12 | 0.24 | 0.28 | 0.16 | 0.19 | 0.14 |
| | CI | 0.05-0.32 | 0.11-0.46 | 0.03-0.28 | 0.11-0.44 | 0.14-0.48 | 0.06-0.33 | 0.08-0.37 | 0.05-0.29 |
| D53 Total Anomalous Pulmonary Venous Return | Cases Rate CI | 5 0.08 0.17-0.23 | 5 0.07 0.01-0.22 | 13 0.19 0.08-0.38 | 17 0.25 0.12-0.46 | 13 0.19 0.08-0.37 | 11 0.16 0.06-0.33 | 11 0.16 0.06-0.33 | 12 0.16 0.07-0.33 |
| E01 Choanal Atresia | Cases | 6 | 10 | 10 | 16 | 6 | 5 | 6 | 14 |
| | Rate | 0.09 | 0.15 | 0.15 | 0.24 | 0.09 | 0.07 | 0.09 | 0.19 |
| | CI | 0.24-0.25 | 0.05-0.33 | 0.05-0.32 | 0.11-0.44 | 0.02-0.23 | 0.02-0.21 | 0.02-0.23 | 0.09-0.37 |
| E06 Agenesis of Lung | Cases | 25 | 44 | 32 | 42 | 49 | 50 | 26 | 14 |
| | Rate | 0.40 | 0.69 | 0.48 | 0.62 | 0.71 | 0.73 | 0.38 | 0.19 |
| | CI | 0.22-0.67 | 0.45-1.00 | 0.29-0.75 | 0.40-0.92 | 0.47-1.01 | 0.49-1.04 | 0.21-0.61 | 0.09-0.37 |
| F01 Cleft Palate | Cases | 39 | 46 | 36 | 43 | 38 | 31 | 27 | 47 |
| | Rate | 0.63 | 0.72 | 0.54 | 0.64 | 0.55 | 0.45 | 0.39 | 0.64 |
| | CI | 0.40-0.95 | 0.47-1.04 | 0.33-0.82 | 0.41-0.93 | 0.35-0.82 | 0.27-0.71 | 0.22-0.63 | 0.43-0.93 |

| CODE/CONDITION (1) | | | | | | | | | |
|---|---------------------|--------------------------|--------------------------|--------------------------|--------------------------|--------------------------|--------------------------|--------------------------|--------------------------|
| | | <u>1986</u> | <u>1987</u> | <u>1988</u> | <u>1989</u> | <u>1990</u> | <u>1991</u> | <u>1992</u> | <u>1995</u> |
| F02 Cleft Lip with and without Cleft Palate | Cases Rate CI | 77 1.25 0.91-1.67 | 80 1.25 0.92-1.66 | 91 1.37 1.03-1.79 | 90 1.33 1.00-1.74 | 97 1.40 1.06-1.81 | 80 1.17 0.86-1.55 | 74 1.07 0.78-1.43 | 94 1.29 0.97-1.67 |
| F08 Pyloric Stenosis | Cases Rate CI | 108 1.76 1.35-2.25 | 135 2.11 1.67-2.63 | 134 2.03 1.60-2.52 | 122 1.81 1.41-2.27 | 116 1.68 1.30-2.12 | 148 2.16 1.73-2.66 | 137 1.98 1.57-2.46 | 148 2.03 1.63-2.50 |
| F09 TE Fistula, or Esophageal Atresia, or both | Cases Rate CI | 19 0.31 0.15-0.54 | 16 0.25 0.11-0.46 | 19 0.28 0.14-0.50 | 18 0.27 0.13-0.48 | 19 0.27 0.14-0.48 | 15 0.22 0.10-0.41 | 14 0.20 0.09-0.39 | 18 0.25 0.12-0.44 |
| F14 Stenosis/Atresia of Duodenum | Cases Rate CI | 5 0.08 0.01-0.23 | 15 0.07 0.01-0.22 | 11 0.16 0.06-0.34 | 10 0.15 0.05-0.32 | 10 0.14 0.05-0.31 | 6 0.09 0.02-0.23 | 13 0.19 0.08-0.37 | 9 0.12 0.04-0.28 |
| F15 Stenosis/Atresia of Small Intestine | Cases Rate CI | 18 0.29 0.14-0.52 | 12 0.18 0.07-0.37 | 13 0.19 0.08-0.38 | 16 0.24 0.11-0.44 | 16 0.23 0.11-0.43 | 9 0.13 0.05-0.29 | 13 0.19 0.08-0.37 | 11 0.15 0.06-0.31 |
| F16 Stenosis/Atresia of Rectum or Anus | Cases Rate CI | 27 0.44 0.25-0.71 | 26 0.40 0.23-0.66 | 27 0.40 0.23-0.66 | 35 0.52 0.32-0.79 | 35 0.51 0.31-0.78 | 38 0.56 0.35-0.83 | 31 0.45 0.27-0.70 | 37 0.51 0.32-0.77 |
| F17 Hirschsprung's Disease | Cases Rate CI | 11 0.17 0.07-0.37 | 10 0.15 0.05-0.33 | 10 0.15 0.05-0.32 | 7 0.03 0.03-0.25 | 13 0.19 0.08-0.37 | 13 0.19 0.08-0.37 | 7 0.10 0.03-0.25 | 16 0.22 0.10-0.41 |

Table 3 Continued Arizona Birth Defects Monitoring Program Incidence Rates^a Per 1,000 Live Births and Fetal Deaths, Arizona

| CODE/CONDITION (1) | | | | | | | | | |
|--|---------------------|------------------------|-------------------------|------------------------|------------------------|------------------------|------------------------|------------------------|------------------------|
| | | <u>1986</u> | <u>1987</u> | <u>1988</u> | <u>1989</u> | <u>1990</u> | <u>1991</u> | <u>1992</u> | <u>1995</u> |
| F18 Malrotation of Intestine | Cases | 10 | 10 | 16 | 14 | 16 | 14 | 10 | 19 |
| | Rate | 0.16 | 0.15 | 0.24 | 0.21 | 0.23 | 0.20 | 0.14 | 0.26 |
| | CI | 0.06-0.35 | 0.05-0.33 | 0.11-0.44 | 0.09-0.40 | 0.11-0.43 | 0.09-0.39 | 0.05-0.31 | 0.13-0.46 |
| F21 Biliary Atresia | Cases | 2 | 1 | 3 | 5 | 4 | 6 | 4 | 3 |
| | Rate | 0.03 | 0.01 | 0.04 | 0.07 | 0.06 | 0.09 | 0.06 | 0.04 |
| | CI | 0.00-0.15 | 0.00-0.11 | 0.00-0.16 | 0.02-0.21 | 0.01-0.18 | 0.02-0.23 | 0.01-0.18 | 0.00-0.15 |
| H01 Renal Agenesis | Cases | 21 | 27 | 23 | 43 | 33 | 37 | 33 | 39 |
| | Rate | 0.34 | 0.42 | 0.34 | 0.64 | 0.48 | 0.54 | 0.48 | 0.54 |
| | CI | 0.18 | 0.24-0.68 | 0.18-0.58 | 0.41-0.93 | 0.29-0.74 | 0.34-0.82 | 0.29-0.74 | 0.34-0.80 |
| H06 Obstruction Kidney/Ureter | Cases | 37 | 71 | 64 | 90 | 94 | 103 | 73 | 108 |
| | Rate | 0.60 | 1.11 | 0.97 | 1.33 | 1.36 | 1.50 | 1.05 | 1.48 |
| | CI | 0.37-0.91 | 0.80-1.50 | 0.68-1.32 | 1.00-1.74 | 1.02-1.76 | 1.15-1.93 | 0.76-1.42 | 1.14-1.89 |
| H09 Bladder or Urethra Obstruction | Cases Rate CI | 8 0.13 0.04-0.30 | 12 0.18 0.07-0.37 | 9 0.13 0.04-0.30 | 7 0.10 0.03-0.25 | 3 0.04 0.00-0.16 | 8 0.12 0.04-0.27 | 7 0.10 0.03-0.25 | 7 0.10 0.03-0.24 |
| J03 Dislocation of Hip | Cases | 87 | 101 | 68 | 91 | 105 | 103 | 66 | 83 |
| | Rate | 1.42 | 1.58 | 1.03 | 1.35 | 1.52 | 1.50 | 0.95 | 1.14 |
| | CI | 1.05-1.86 | 1.20-2.03 | 1.20-2.03 | 1.01-1.76 | 1.16-1.76 | 1.15-1.93 | 0.68-1.30 | 0.84-1.50 |
| J51 Complete absence of upper or lower limb | Cases Rate CI | 2 0.03 0.00-0.15 | 0 0.00 0.00-0.00 | 1 0.01 0.00-0.11 | 3 0.04 0.00-0.16 | 3 0.04 0.00-0.16 | 2 0.03 0.00-0.14 | 1 0.01 0.00-0.11 | 2 0.03 0.00-0.13 |

Table 3 Continued Arizona Birth Defects Monitoring Program Incidence Rates^a Per 1,000 Live Births and Fetal Deaths, Arizona

Table 3 Continued Arizona Birth Defects Monitoring Program Incidence Rates^a Per 1,000 Live Births and Fetal Deaths, Arizona

| CODE/CONDITION (1) | | | | | | | | | | |
|------------------------------------|---------------------|-------------------------|-------------------------|-------------------------|-------------------------|-------------------------|-------------------------|-------------------------|-------------------------|--|
| | | <u>1986</u> | <u>1987</u> | <u>1988</u> | <u>1989</u> | <u>1990</u> | <u>1991</u> | <u>1992</u> | <u>1995</u> | |
| J52 Phocomelia of limb | Cases | 3 | 2 | 2 | 1 | 1 | 1 | 0 | 0 | |
| | Rate | 0.04 | 0.03 | 0.03 | 0.01 | 0.01 | 0.01 | 0.00 | 0.00 | |
| | CI | 0.00-0.18 | 0.00-0.14 | 0.00-0.14 | 0.00-0.25 | 0.00-0.11 | 0.00-0.11 | 0.00-0.00 | 0.00-0.00 | |
| K05 Amniotic Bands | Cases | 4 | 4 | 9 | 8 | 14 | 10 | 8 | 12 | |
| | Rate | 0.06 | 0.06 | 0.14 | 0.11 | 0.20 | 0.15 | 0.12 | 0.16 | |
| | CI | 0.01-0.20 | 0.01-0.19 | 0.05-0.32 | 0.04-0.28 | 0.09-0.39 | 0.05-0.31 | 0.04-0.27 | 0.07-0.33 | |
| N01 Diaphragmatic Hernia | Cases | 13 | 18 | 20 | 23 | 28 | 23 | 13 | 20 | |
| | Rate | 0.21 | 0.28 | 0.30 | 0.34 | 0.40 | 0.34 | 0.19 | 0.27 | |
| | CI | 0.09-0.41 | 0.13-0.50 | 0.15-0.52 | 0.19-0.57 | 0.23-0.65 | 0.18-0.56 | 0.08-0.37 | 0.14-0.48 | |
| N02 Omphalocele | Cases | 10 | 14 | 17 | 10 | 21 | 21 | 10 | 14 | |
| | Rate | 0.16 | 0.21 | 0.25 | 0.15 | 0.30 | 0.31 | 0.14 | 0.19 | |
| | CI | 0.06-0.35 | 0.09-0.42 | 0.12-0.46 | 0.05-0.32 | 0.16-0.52 | 0.16-0.53 | 0.05-0.31 | 0.09-0.37 | |
| N04 Gastroschisis | Cases | 19 | 18 | 19 | 19 | 21 | 36 | 27 | 27 | |
| | Rate | 0.31 | 0.28 | 0.28 | 0.28 | 0.30 | 0.53 | 0.39 | 0.37 | |
| | CI | 0.15-0.54 | 0.13-0.50 | 0.14-0.50 | 0.14-0.50 | 0.16-0.52 | 0.33-0.80 | 0.22-0.63 | 0.21-0.60 | |
| R01 Down Syndrome (Trisomy 21) | Cases Rate CI | 64 1.04 0.73-1.43 | 61 0.95 0.67-1.32 | 74 1.12 0.81-1.50 | 66 0.98 0.70-1.33 | 73 1.05 0.76-1.42 | 84 1.23 0.91-1.62 | 87 1.26 0.94-1.65 | 90 1.23 0.92-1.61 | |
| R02 Patau Syndrome (Trisomy 13) | Cases Rate CI | 9 0.14 0.05-0.32 | 4 0.06 0.01-0.19 | 3 0.04 0.00-0.16 | 4 0.06 0.01-0.19 | 11 0.16 0.06-0.33 | 6 0.09 0.02-0.23 | 15 0.22 0.10-0.41 | 8 0.11 0.03-0.26 | |

Table 3 Continued Arizona Birth Defects Monitoring Program Incidence Rates Per 1,000 Live Births and Fetal Deaths¹ Arizona, 1995

| CODE/CONDITION (1) | | | | | | | | | | |
|--------------------------------------|---------------------|-------------------------|-------------------------|-------------------------|-------------------------|-------------------------|-------------------------|-------------------------|-------------------------|--|
| | | <u>1986</u> | <u>1987</u> | <u>1988</u> | <u>1989</u> | <u>1990</u> | <u>1991</u> | <u>1992</u> | <u>1995</u> | |
| R03 Edwards Syndrome (Trisomy 18) | Cases Rate CI | 11 0.17 0.07-0.37 | 17 0.26 0.12-0.48 | 13 0.19 0.08-0.38 | 10 0.15 0.05-0.32 | 15 0.22 0.10-0.41 | 13 0.19 0.08-0.37 | 12 0.17 0.07-0.35 | 18 0.25 0.12-0.44 | |
| S02 Fetal Alcohol Syndrome | Cases Rate CI | 9 0.14 0.05-0.32 | 25 0.39 0.21-0.64 | 12 0.18 0.07-0.36 | 21 0.31 0.16-0.53 | 22 0.32 0.17-0.54 | 27 0.39 0.23-0.64 | 33 0.48 0.29-0.74 | 27 0.37 0.21-0.60 | |

(1) See appendix for explanation of the codes/conditions

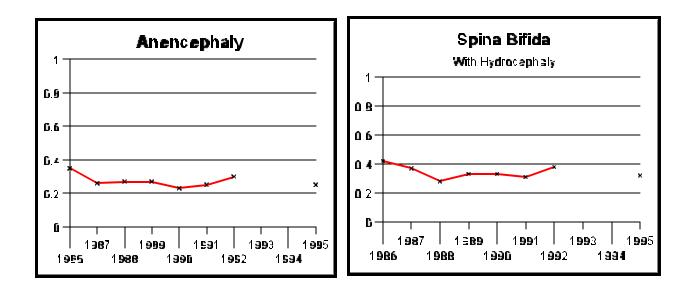
CI = Approximate 99% confidence intervals. "Cases" are the number of live born and still born infants >= 20 weeks.

^a The rates are calculated as the number of live born and still born cases of each defect divided by the denominators consisting of the total live births and still births as follows:

Denominators -

1986 = 61,203; 1987 = 63,742; 1988 = 65,981; 1989 = 67,498; 1990 = 69,245; 1991 = 68,449; 1992 = 69,202; 1995 = 72,883.

Figure 2. Trends of Selected Congenital Anomalies: Incident Rates (Live Born and Still Born Cases **Per 1,000 Live Births & Fetal Deaths**), Arizona^a



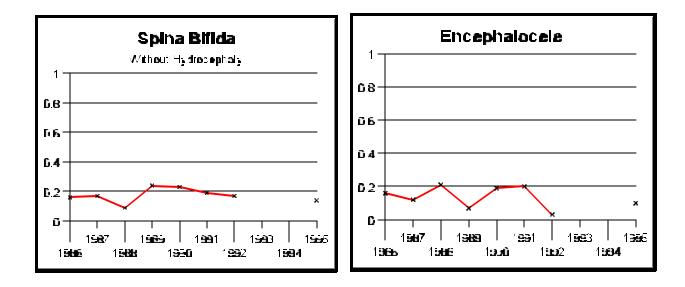


Figure 2 Continued Trends of Selected Congenital Anomalies: Incidence Rates (Live Born and Still Born Cases Per 1,000 Live Births & Fetal Deaths), Arizona^a

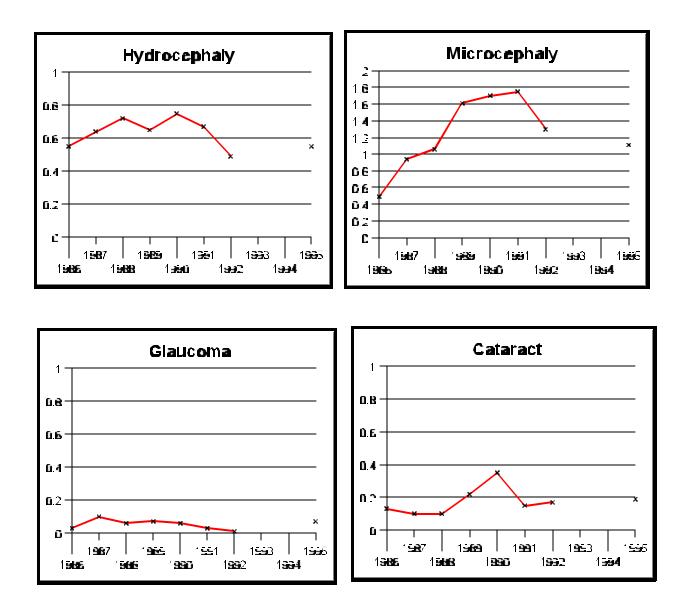


Figure 2 Continued Trends of Selected Congenital Anomalies: Incidence Rates (Live Born and Still Born Cases Per 1,000 Live Births & Fetal Deaths), Arizona^a

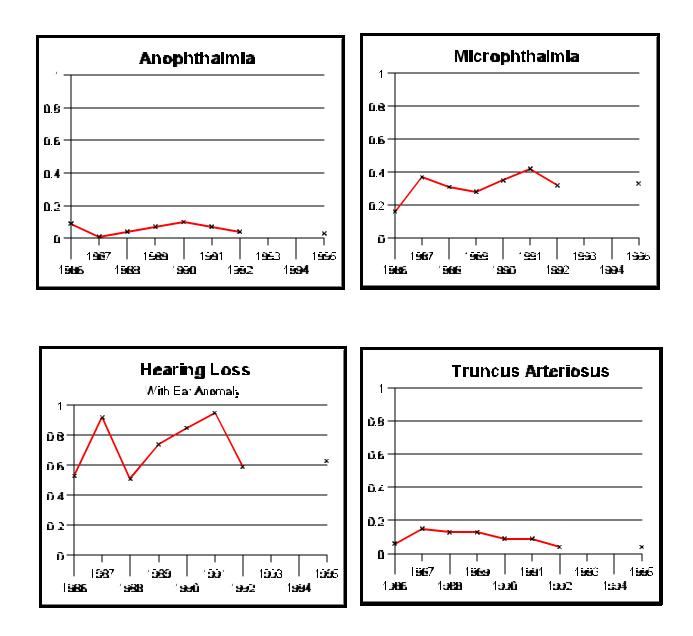
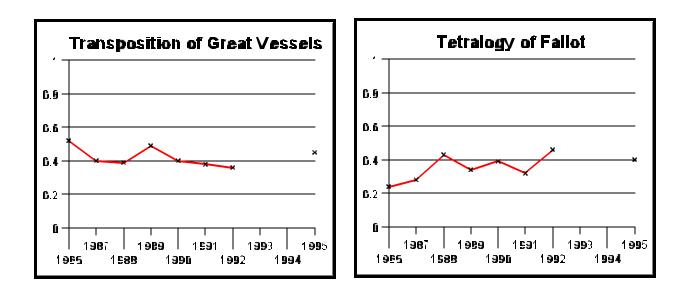


Figure 2 Continued Trends of Selected Congenital Anomalies: Incidence Rates (Live Born and Still Born Cases Per 1,000 Live Births & Fetal Deaths), Arizona^a



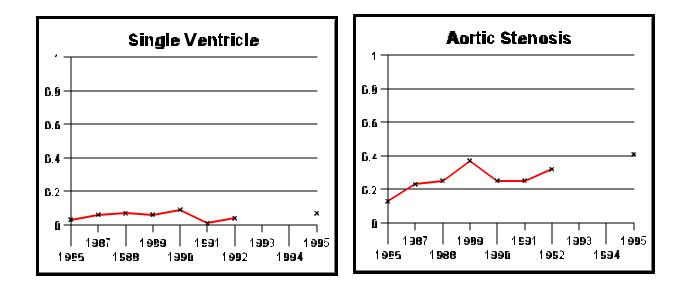


Figure 2 Continued Trends of Selected Congenital Anomalies: Incidence Rates (Live Born and Still Born Cases Per 1,000 Live Births & Fetal Deaths), Arizona^a

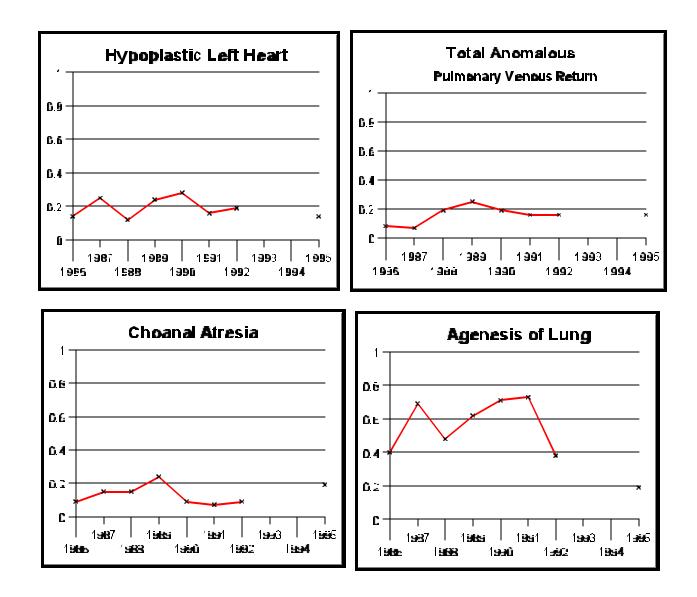


Figure 2 Continued Trends of Selected Congenital Anomalies: Incidence Rates (Live Born and Still Born Cases Per 1,000 Live Births & Fetal Deaths), Arizona^a

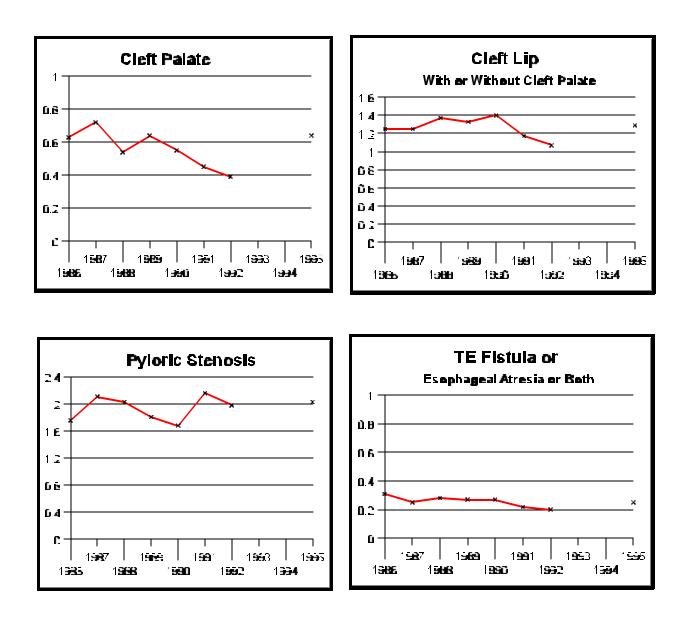


Figure 2 Continued Trends of Selected Congenital Anomalies: Incidence Rates (Live Born and Still born Cases Per 1,000 Live Births & Fetal Deaths), Arizona^a

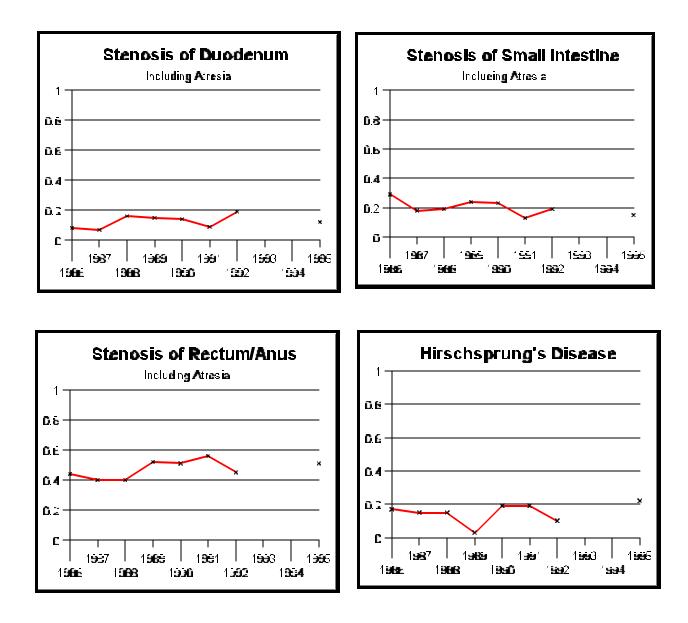


Figure 2 Continued Trends of Selected Congenital Anomalies: Incidence Rates (Live Born and Still Born Cases Per 1,000 Live Births & Fetal Deaths), Arizona^a

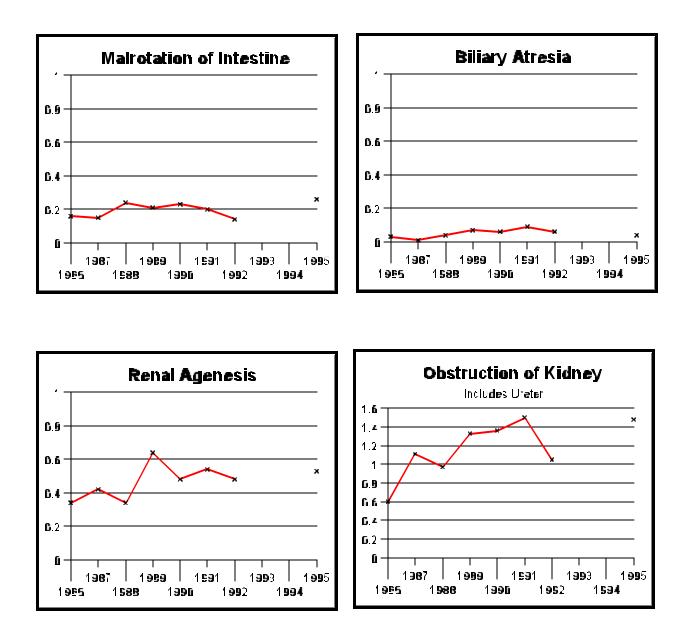


Figure 2 Continued Trends of Selected Congenital Anomalies: Incidence Rates (Live Born and Still Born Cases Per 1,000 Live Births & Fetal Deaths), Arizona^a

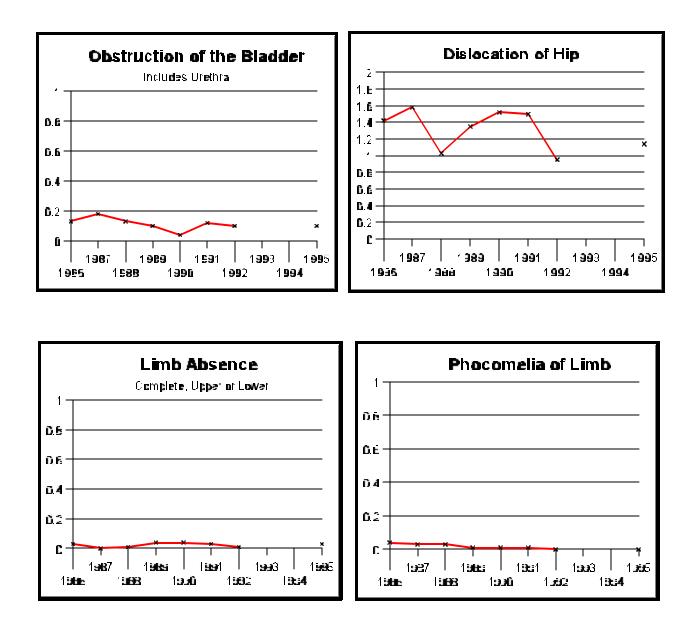


Figure 2 Continued Trends of Selected Congenital Anomalies: Incidence Rates (Live Born and Still Born Cases Per 1,000 Live Births & Fetal Deaths), Arizona^a

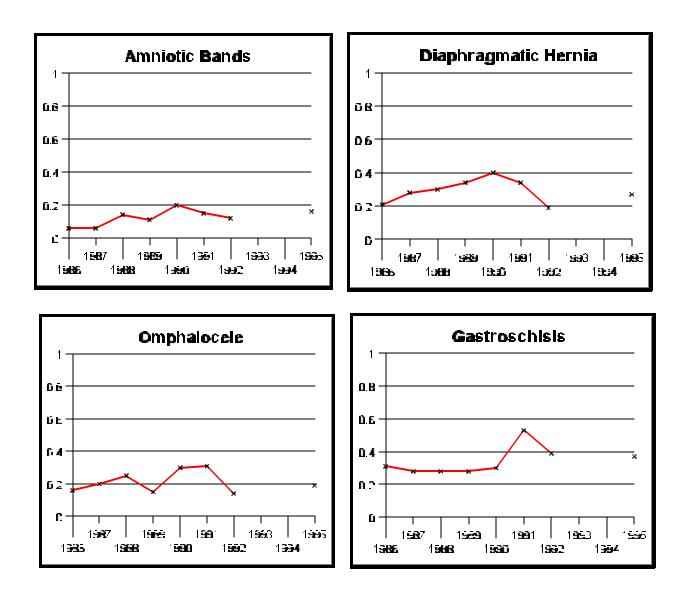
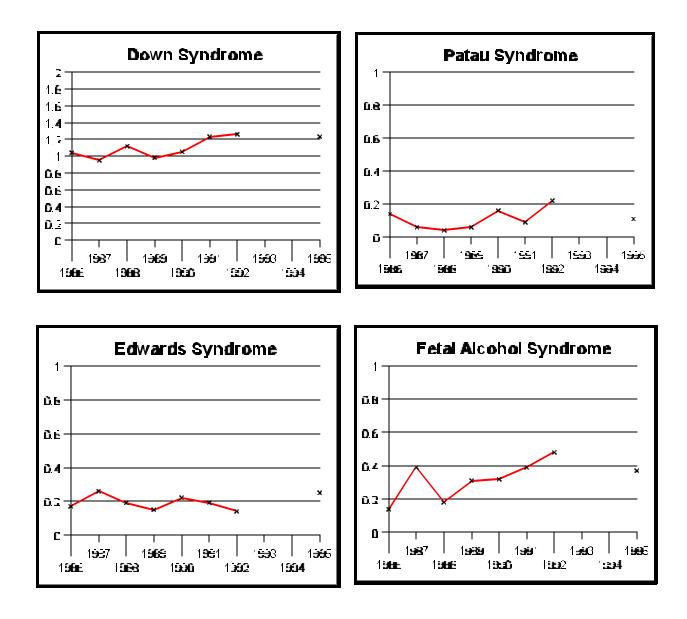


Figure 2 Continued Trends of Selected Congenital Anomalies: Incidence Rates (Live Born and Still Born Cases Per 1,000 Live Births & Fetal Deaths), Arizona^a



RACE/ETHNICITY

Race and ethnicity categories were determined from the mother's race and Hispanic origin portion of the child's birth certificate. The Hispanic category consists of mothers who answered 'White' to race and 'Hispanic' to the Hispanic origin question. The remaining race categories are White Non-Hispanic, Black, Native American, and Other. The graphs do not show the rates for all of the race/ethnic groups due to the small number of cases of specific birth defects among the subgroups. Table 1 shows the counts used for the calculation of the rates.

Spina Bifida was the most common neural tube defect (NTD) among all races. Rates of Spina Bifida were highest among Hispanics, but is not statistically significant (Figure 3). Many studies have documented that Hispanics have higher rates of Spina Bifida compared to Whites. The same pattern was found in the rates of Anencephaly. The literature also suggests that Blacks experience lower rates of Spina Bifida and Anencephaly compared to Whites; however, rate comparisons of NTDs were limited to White and Hispanic due to small number of cases occurring among other races.

Unlike in previous years, there appears to be a reversal in the rates of abdominal wall defects among Hispanics and Whites, with the higher rate found in Whites (Figure 4). Examining specific defects, the rates for both Gastroschisis and for Omphalocele are higher among Whites, relative to Hispanics. These patterns however, are not statistically significant. Again, rate comparisons among other races was not possible due to small numbers.

Down Syndrome (Trisomy 21) rates were highest among Blacks, followed by Hispanics, Whites and Native Americans (Figure 5). These differences however, are not statistically significant.

Microcephaly rates are highest among Blacks (Figure 6), while Hispanics, Blacks, followed by Whites had the highest rates of pyloric stenosis (Figure 7). Statistical analysis of these rates indicated that these are statistically significant.

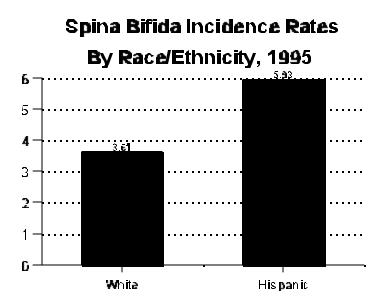


Figure 3. Spina Bifida Incidence Rates (Live Born and Still Born Cases Per 10,000 Live Births and Fetal Deaths) by Race/Ethnicity, 1995

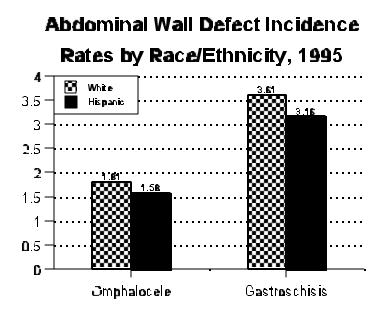


Figure 4. Abdominal Wall Defect Incidence Rates (Live Born and Still Born Cases Per 10,000 Live Births and Fetal Deaths) by Race/Ethnicity, 1995

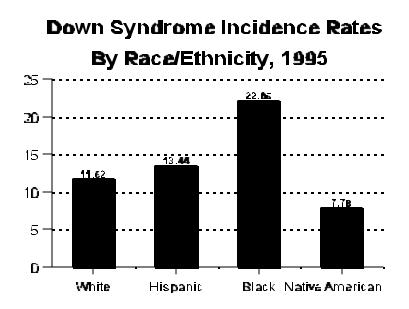


Figure 5. Down Syndrome Incidence Rates (Live Born and Still Born Cases Per 10,000 Live Births and Fetal Deaths) by Race/Ethnicity, 1995

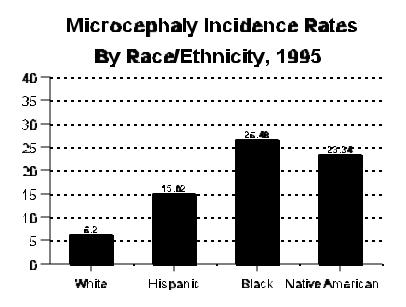


Figure 6. Microcephaly Incidence Rates (Live Born and Still Born Cases Per 10,000 Live Births and Fetal Deaths) by Race/Ethnicity, 1992

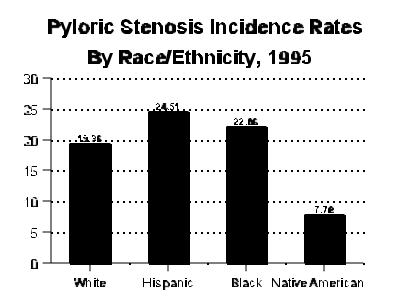


Figure 7. Pyloric Stenosis Incidence Rates (Live Born and Still Born Cases Per 10,000 Live Births and Fetal Deaths) by Race/Ethnicity, 1995

MATERNAL AGE

Maternal age was divided into five age groups. Observed rates of the "44 selected" congenital anomalies were highest among women 35 years of age and older, followed by the less than 20 age group (Figure 8). Down syndrome (Trisomy 21) rates increased with maternal age (Figure 9). In contrast, rates for gastroschisis decreased as maternal age increased (Figure 10).

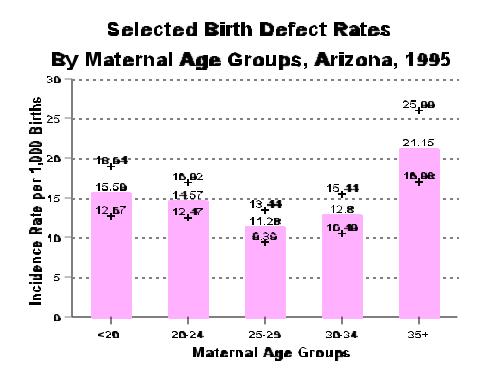
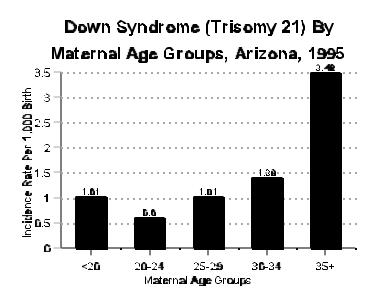
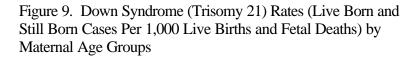


Figure 8. Incidence Rates (Live Born and Still Born Cases Per 1,000 Live Births and Fetal Deaths) for the 44 Selected Defects Listed on Table 1. The + sign indicates the 99% confidence bounds.





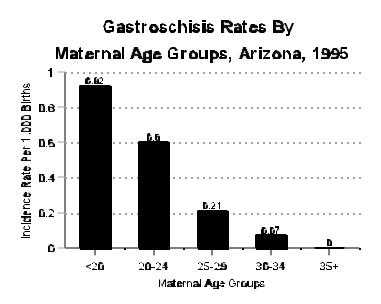


Figure 10. Gastroschisis Rates (Live Born and Still Born Cases Per 1,000 Live Births and Fetal Deaths) by Maternal Age Groups

COUNTY PROFILES

Using County Data

The Arizona Birth Defect Monitoring Program (ABDMP) collects birth defect information from all of Arizona's 15 counties. Multiple years are used to give sufficient data to derive statistically stable measures at the county level. Birth defect data from live births are analyzed in this section.

Dealing With Small Numbers

Analysis of county data is difficult because of normal fluctuations in rates seen in small populations. When dealing with small numbers, it is normal to see fluctuations over time. With rate fluctuations we may see the appearance of birth defects clusters. Most often this is a statistical anomaly. In the rare case that a cluster results from a teratogen a dramatic increase on the scale of 10-fold or greater is usually seen.¹³ Another concern with small numbers is protecting a person and their family's confidentiality. Thus, all county level data are aggregated. Incidence rates and confidence intervals are presented when there are 10 or more cases. Z-tests were used to test for the equivalence between the county rates with the state rate.

Birth Defects by County

The following tables present birth defects by county of mothers' residence. Cases were aggregated for the years 1986 through 1992 and 1995 to provide large enough numbers for analysis. Table 4 shows the total number of 44 selected congenital anomalies for each Arizona county. Gila county had the highest rate of congenital anomalies, whereas Greenlee and La Paz counties had the lowest reported birth defects rates. Statistical analysis indicate that the overall birth defect rates of Gila is significantly higher than the state rate at the 0.01 level. Table 5 examines the 44 selected anomalies by race and county. For Whites, Maricopa county has the highest rate, followed by Yavapai county. For Hispanics, Yavapai county has the highest rate of congenital anomalies and Pima county has the highest rate for Blacks. Graham and Gila counties have the highest rates for Native Americans. The rates for each race/ethnicity group for each county is compared with that of the state rate for that race/ethnic group. Statistical analysis of the data indicate that the Apache county rates for Whites is significantly lower than the state rate for whites at the 99 percent level. In contrast, the data show that the Gila county rates for Native Americans is significantly higher at the 0.01 level than the state rate for Native Americans. Other county rates for the other race/ethnic groupings were not significantly different from the state rate for that race/ethnic group.

| COUNTY | CASES 1986-1992, 1995 | RATE | 99% CONFIDENCE INTERVAL |
|------------|-----------------------------|-------|----------------------------|
| Arizona | 7146 | 13.36 | 12.96-13.78 |
| Apache | 192 | 13.94 | 11.48-16.76 |
| Cochise | 156 | 11.68 | 9.41-14.32 |
| Coconino | 216 | 14.03 | 11.69-16.69 |
| Gila | 86 | 16.31 | 12.13-21.42 |
| Graham | 43 | 12.62 | 08.21-18.50 |
| Greenlee | 7 | - | - |
| Maricopa | 4,186 | 13.34 | 12.82-13.89 |
| Mohave | 124 | 11.05 | 08.66-13.88 |
| Navajo | 241 | 15.89 | 13.37-18.72 |
| Pima | 1176 | 13.17 | 12.20-14.20 |
| Pinal | 229 | 13.71 | 11.49-16.23 |
| Santa Cruz | 72 | 12.70 | 09.17-17.10 |
| Yavapai | 132 | 13.07 | 10.32-16.30 |
| Yuma | 271 | 14.12 | 12.00-16.49 |
| La Paz | 15 | 9.68 | 04.43-18.20 |

Table 4 Selected Birth Defect Incidence Rates by County 1986-1992, 1995 Incidence Rates (Live Born Cases Per 1,000 Live Births)

44 selected birth defects (see Table 1); - =Insufficient cases for rate and confidence interval calculations

| Incidence Rates (Live Born Cases Per 1,000 Live Births) | | | | | |
|---|----------------------|----------------------|----------------------|----------------------|---------------------|
| COUNTY | WHITE | HISPANIC | BLACK | NATIVE AMERICAN | OTHER |
| | Rate 99% C.I. | Rate 99% C.I. | Rate 99% C.I. | Rate 99% C.I. | Rate 99% C.I. |
| Arizona | 12.5 11.96-13.02 | 13.90 13.14-14.68 | 12.12 10.37-14.08 | 18.03 16.47-19.68 | 10.92 8.07-14.12 |
| Apache | 1.92 0.85-3.68 | - | - | 14.99 12.23-18.17 | - |
| Cochise | 12.00 8.79-15.98 | 11.79 8.23-16.31 | 9.08 3.54-18.85 | - | - |
| Coconino | 9.78 6.85-13.50 | 13.20 6.62-21.59 | - | 17.54 13.74-22.03 | - |
| Gila | 10.42 5.82-17.10 | 10.96 4.68-21.54 | | 27.74 18.23-40.31 | |
| Graham | 9.66 4.98-16.76 | - | - | 29.94 13.70-56.30 | - |
| Greenlee | - | - | - | - | - |
| Maricopa | 12.86 12.21-13.54 | 14.34 13.31-15.42 | 11.90 9.82-14.26 | 18.90 15.35-23.00 | 9.69 6.60-13.68 |
| Mohave | 10.77 8.21-13.86 | 11.66 5.34-21.94 | - | - | - |
| Navajo | 11.77 8.07-17.25 | 14.55 6.44-27.94 | - | 17.76 14.49-21.53 | - |
| Pima | 12.21 10.89-13.59 | 13.53 11.97-15.23 | 14.94 10.46-20.62 | 19.60 14.22-26.29 | 12.49 6.57-21.41 |
| Pinal | 12.01 10.91-13.62 | 13.27 9.81-17.51 | - | 21.91 14.86-31.05 | - |
| Santa Cruz | - | 13.24 9.41-18.06 | - | - | - |
| Yavapai | 12.84 9.85-16.42 | 15.67 8.51-26.25 | - | - | - |
| Yuma | 12.22 9.00-16.18 | 15.04 12.24-18.27 | - | - | - |
| La Paz | - | - | - | - | - |

Table 5 – Selected Birth Defects by Race/Ethnicity by County, 1986-1992, 1995 Incidence Rates (Live Born Cases Per 1,000 Live Births)

- =Insufficient cases for rate and confidence interval calculations.

SENTINEL DEFECTS

Tables 6-10 look at the following sentinel defects: chromosomal defects, oral clefts, neural tube defects, abdominal wall defects, and heart defects. These defects were chosen because of their significant public health impact.

Chromosomal Defects

In this section (Table 6) of the report, chromosomal defects refers to Down Syndrome, Patau syndrome, and Edwards syndrome. Chromosomal abnormalities include either missing or extra genetic components that result in various levels of abnormal physical features, structural defects, and mental retardation. The most common chromosomal defects is Down Syndrome. We also know that the risk of a trisomy affected pregnancy increases with maternal age; however, this risk is still relatively low. Recent research also suggests that about 20 percent of instances of Down Syndrome

are paternal in origin. Table 6 shows that rates for chromosomal defects are highest for Gila county (2.28 per 1,000 live births), followed by Navajo county at 1.91 per 1,000 live births. The lowest rates are in Greenlee, La Paz, Graham, Santa Cruz and Mohave counties. A comparison of the county rates with the state rate for chromosomal defects indicate that there are no significant differences between the county rates and that of the state.

Oral Clefts

Table 7 presents information on cleft lip and cleft palate. Cleft palate is a failure of the palate to fuse properly, forming a grooved fissure in the roof of the mouth. Cleft lip is a failure of the maxillary and median nasal processes to fuse, forming a fissure in the lip. Babies born with oral clefts require corrective surgery, and may have feeding problems. Mothers who smoke 20 or more cigarettes a day are more than twice as likely to have a baby born with cleft lip and/or cleft palate.¹⁴ Apache county, followed by Gila county have the highest rates for oral clefts at 3.05 and 2.84 per 1,000 live births respectively. Greenlee, La Paz, Graham and Maricopa counties, on the other hand have the lowest rates. The state rate for oral clefts is 1.76 per 1,000 live births from 1986 to 1992 and 1995. Results of the z-tests comparing the oral clefts rates of the counties with the state show that there are no statistical differences between these rates. A map showing the incidence rates of oral clefts by county for 1986 to 1992 and 1995 are in Figure 10. In Figure 11 is a map displaying the distribution of oral cleft cases in 1995. The distribution of cleft palate cases and the distribution of cleft lip with and without cleft palate cases are also presented. It can be seen from the maps that the cases appear to be widely dispersed across the state (Figure 11).

Neural Tube Defects

Anencephaly, spina bifida, and encephalocele make up the neural tube defects (NTDs) presented in Table 8. The two major NTDs are anencephaly and spina bifida. Anencephaly is an absence of the skull, with cerebral hemispheres reduced or completely missing. Spina bifida is a defective closure of the bony encasement of the spinal cord, through which the cord and meninges may or may not protrude. Women who take multivitamins and/or eat a diet rich in folate before conception and during the first trimester^{16,17} can significantly reduce their risk of an NTD affected pregnancy. The data show that rate for neural tube defect for the state is 0.71 per 1,000 live births. A comparison between the county rates and the state rate indicate that Navajo county has the highest rate for neural tube defect at 1.25 per 1,000 live births, but is not statistically different from the state rate.

Abdominal Wall Defects

This category includes omphalocele and gastroschisis (Table 9). Gastroschisis is a congenital opening of the abdominal wall, often with protrusion of the intestines. Omphalocele is a membrane-covered protrusion of an abdominal organ through the abdominal wall at the umbilicus. According to a recent study, young mothers are 4 times as likely as women in their late 20s to have a child with gastroschisis.¹⁵ Other risk factors for gastroschisis are maternal use of cocaine, aspirin, amphetamines, and exposure to solvents. Table 9 presents the incidence rate for the state at 0.50 per 1,000 live births. Mohave county has the highest incidence rate for abdominal wall defects. There are no statistical differences between the county rates and the state rate for abdominal wall defects.

Heart Defects

This category includes truncus Arteriosus, transposition of great vessels, Tetralogy of Fallot, single ventricle, aortic stenosis, hypoplastic left heart, and total anomalous pulmonary venous (Table 10). Table 10 shows that state rate for heart defects is 1.44 per 1,000 live births. Navajo county and Gila county have the highest rate for heart defects at 2.11 and 2.09 per 1,000 live births. The county rates for heart defects were found not to be statistically different from the rate at the state level.

| COUNTY | CASES 1986-1992 & 1995 | RATE | 99% CONFIDENCE INTERVAL |
|------------|------------------------------|------|----------------------------|
| Arizona | 707 | 1.32 | 1.20-1.46 |
| Apache | 25 | 1.82 | 1.01-2.98 |
| Cochise | 22 | 1.65 | 0.88-2.79 |
| Coconino | 21 | 1.36 | 0.72-2.34 |
| Gila | 12 | 2.28 | 0.93-4.59 |
| Graham | 6 | - | - |
| Greenlee | 0 | - | - |
| Maricopa | 401 | 1.28 | 1.12-1.45 |
| Mohave | 13 | 1.16 | 0.49-2.28 |
| Navajo | 29 | 1.91 | 1.12-3.03 |
| Pima | 112 | 1.25 | 0.97-1.59 |
| Pinal | 22 | 1.32 | 0.70-2.23 |
| Santa Cruz | 7 | - | - |
| Yavapai | 13 | 1.29 | 0.55-2.53 |
| Yuma | 21 | 1.09 | 0.58-1.88 |
| La Paz | 3 | - | - |

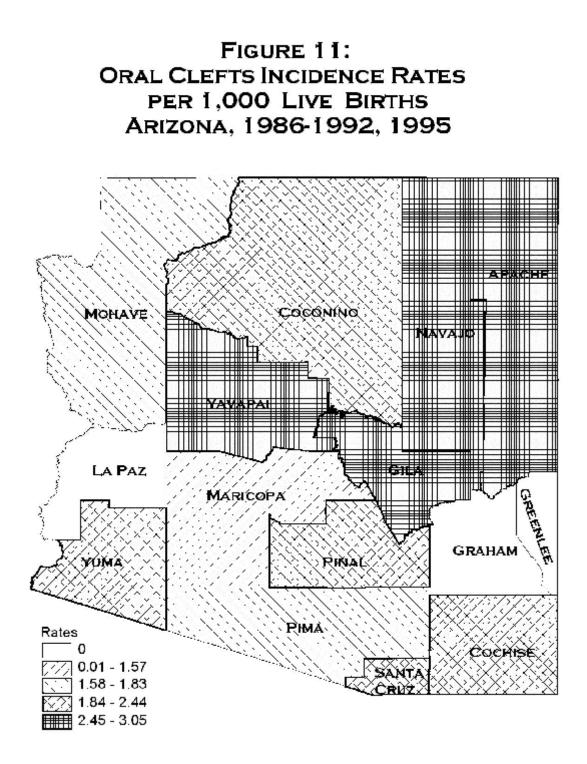
Table 6 Chromosomal Defects - Rates by County 1986-1992, 1995 Incidence Rate (Live Born Cases per 1,000 Live Births)

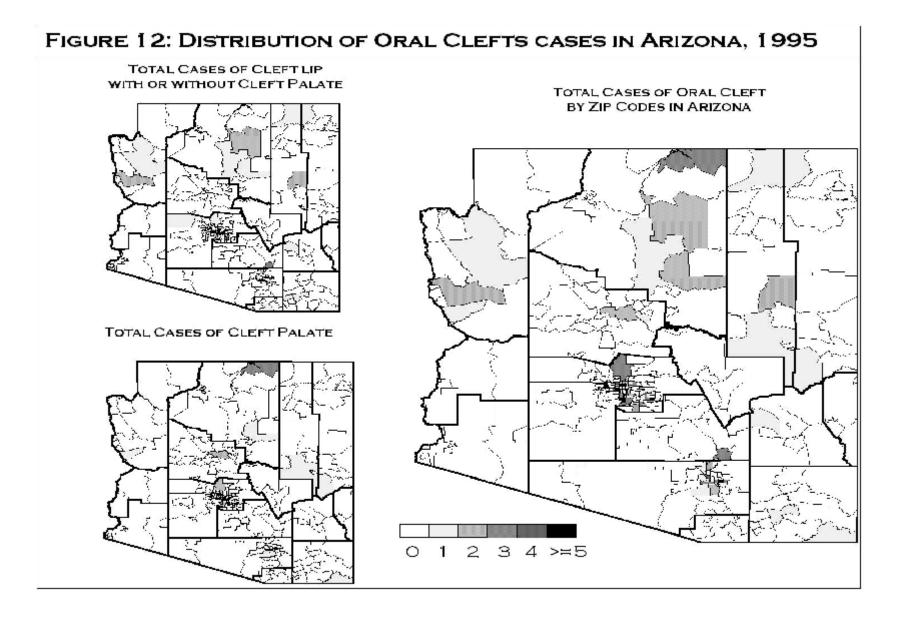
Chromosomal defects include three-digit codes R01, R02, R03 (see Table 1); - =Insufficient cases for rate and confidence interval calculations.

| COUNTY | CASES 1986-1992 & 1995 | RATE | 99% CONFIDENCE INTERVAL |
|------------|------------------------------|------|----------------------------|
| Arizona | 944 | 1.76 | 1.62-1.92 |
| Apache | 42 | 3.05 | 1.97-4.49 |
| Cochise | 28 | 2.10 | 1.21-3.35 |
| Coconino | 37 | 2.40 | 1.51-3.62 |
| Gila | 15 | 2.84 | 1.30-5.35 |
| Graham | 9 | - | - |
| Greenlee | 0 | - | - |
| Maricopa | 494 | 1.57 | 1.40-1.77 |
| Mohave | 19 | 1.69 | 0.86-2.98 |
| Navajo | 45 | 2.97 | 1.95-4.31 |
| Pima | 150 | 1.68 | 1.35-2.07 |
| Pinal | 32 | 1.92 | 1.15-2.98 |
| Santa Cruz | 12 | 2.12 | 0.87-4.27 |
| Yavapai | 23 | 2.28 | 1.24-3.81 |
| Yuma | 36 | 1.88 | 1.17-2.84 |
| La Paz | 2 | - | - |

Table 7Oral Clefts - Rates by County 1986-1992, 1995Incidence Rates (Live Born Cases Per 1,000 Live Births)

Oral Clefts include three-digit codes F01 & F02 (see Table 1); - =Insufficient cases for rate and confidence interval calculations.





Page 45

| COUNTY | CASES 1986-1992 & 1995 | RATE | 99% CONFIDENCE INTERVAL |
|------------|------------------------------|------|----------------------------|
| Arizona | 382 | 0.71 | 0.62-0.81 |
| Apache | 9 | - | - |
| Cochise | 6 | - | - |
| Coconino | 6 | - | - |
| Gila | 6 | - | - |
| Graham | 2 | - | - |
| Greenlee | 0 | - | - |
| Maricopa | 228 | 0.73 | 0.61-0.86 |
| Mohave | 7 | - | - |
| Navajo | 19 | 1.25 | 0.63-2.20 |
| Pima | 55 | 0.62 | 0.42-0.86 |
| Pinal | 9 | - | - |
| Santa Cruz | 6 | - | - |
| Yavapai | 8 | - | - |
| Yuma | 18 | 0.94 | 0.46-1.67 |
| La Paz | 3 | - | - |

| Table 8 |
|---|
| Neural Tube Defects - Rates by County 1986-1992, 1995 |
| Incidence Rates (Live Born Cases Per 1,000 Live Births) |

Neural Tube defects include three-digit codes A01, A02, A03 & A13. (see Table 1); - =Insufficient cases for rate and confidence interval calculations.

| COUNTY | CASES 1986-1992 & 1995 | RATE | 99% CONFIDENCE INTERVAL |
|------------|------------------------------|------|----------------------------|
| Arizona | 266 | 0.50 | 0.42-0.58 |
| Apache | 3 | - | - |
| Cochise | 3 | - | - |
| Coconino | 6 | - | - |
| Gila | 3 | - | - |
| Graham | 2 | - | - |
| Greenlee | 0 | - | - |
| Maricopa | 150 | 0.48 | 0.38-0.59 |
| Mohave | 10 | 0.89 | 0.33-1.91 |
| Navajo | 7 | - | - |
| Pima | 57 | 0.64 | 0.44-0.89 |
| Pinal | 7 | - | - |
| Santa Cruz | 2 | - | - |
| Yavapai | 6 | - | - |
| Yuma | 10 | 0.52 | 0.19-1.12 |
| La Paz | 0 | - | - |

Table 9 Abdominal Wall Defects - Rates by County 1986-1992, 1995 Incidence Rates (Live Born Cases Per 1,000 Live Births)

Abdominal Wall defects include three-digit codes N02 & N04 (see Table 1); - =Insufficient cases for rate and confidence interval calculations.

| COUNTY | CASES 1986-1992, & 1995 | RATE | 99% CONFIDENCE INTERVAL |
|------------|-------------------------------|------|----------------------------|
| Arizona | 773 | 1.44 | 1.31-1.59 |
| Apache | 16 | 1.16 | 0.55-2.14 |
| Cochise | 18 | 1.35 | 0.67-2.41 |
| Coconino | 19 | 1.23 | 0.62-2.17 |
| Gila | 11 | 2.09 | 0.81-4.33 |
| Graham | 3 | - | - |
| Greenlee | 2 | - | - |
| Maricopa | 454 | 1.48 | 1.28-1.63 |
| Mohave | 10 | 0.89 | 0.33-1.91 |
| Navajo | 32 | 2.11 | 1.27-3.28 |
| Pima | 136 | 1.52 | 1.21-1.89 |
| Pinal | 20 | 1.20 | 0.62-2.08 |
| Santa Cruz | 7 | - | - |
| Yavapai | 18 | 1.78 | 0.88-3.18 |
| Yuma | 27 | 1.41 | 0.81-2.27 |
| La Paz | 0 | - | - |

Table 10Heart Defects - Rates by County 1986-1992, 1995Incidence Rates (Live Born Cases Per 1,000 Live Births)

Heart defects include three-digit codes D01, D02, D03, D04, D51, D52 & D53 (see Table 1); - =Insufficient cases for rate and confidence interval calculations.

Conditions Included in the Figures

A general listing of all conditions used to establish the rates shown in the figures in this report is shown below. Some specific inclusions and exclusions are not listed. As mentioned above, ABDMP collects data on 140 conditions or variations of conditions. The conditions listed below include over 99% of all cases reported through ABDMP.

| BPA 3-Digit Code* | General Code Descriptor |
|-------------------|--|
| 740 - 759 | "Congenital Anomalies" Including but not limited to: |
| 740 | Anencephaly and similar anomalies |
| 741 | Spina Bifida |
| 742 | Other Anomalies of the Nervous System |
| 743 | Anomalies of the eye |
| 744 | Anomalies of the ear, face, and neck |
| 745 | Certain anomalies of the heart |
| 746 | Other anomalies of the heart |
| 747 | Anomalies of the circulatory system |
| 748 | Anomalies of the respiratory system |
| 749 | Cleft palate and cleft lip |
| 750 | Other anomalies of the upper alimentary tract |
| 751 | Anomalies of the digestive system |
| 752 | Anomalies of the genital organs |
| 753 | Anomalies of the urinary system |
| 754 | Certain musculoskeletal deformities |
| 755 | Other anomalies of limbs |
| 756 | Other musculoskeletal anomalies |
| 757 | Congenital anomalies of the integument |
| 758 | Chromosomal anomalies |
| 759 | Other and unspecified anomalies |
| ICD-9-CM Code** | |
| 658.80-658.83 | Amniotic bands |
| 760.71 | Fetal alcohol syndrome |

* British Pediatric Association Classification of Diseases

** International Classification of Disease - 9th Edition, Clinical Modification

Conditions (Composite Categories) Shown in the Tables

A listing of the conditions analyzed in the Tables contained in this report is shown below.

The 44 conditions listed here can be described almost completely by codes created by the Centers for Disease Control's Metropolitan Atlanta Congenital Defects Program (MACDP). These codes are listed in the left below, with exceptions noted. On the right below are the corresponding British Pediatric Association (BPA) Classification of Diseases codes.

In the Tables, a case is listed only once in each MACDP code category, even when it has more than one diagnosis from the category.

| MACDP | Condition | BPA Code | | | | |
|----------------|-----------------------------------|----------|---------|---------|--|--|
| <u>CENTRAI</u> | CENTRAL NERVOUS SYSTEM | | | | | |
| A01 | Anencephaly | 740.00 | 740.02 | 740.03 | | |
| | | 740.08 | 740.10 | 740.20 | | |
| | | 740.21 | 740.29 | | | |
| A02 | Spina Bifida with Hydrocephaly | 741.00 | 741.01 | 741.02 | | |
| | | 741.03 | 741.04 | 741.05 | | |
| | | 741.06 | 741.07 | 741.08 | | |
| | | 741.09 | 741.085 | 741.086 | | |
| | | 741.087 | | | | |
| A03 | Spina Bifida without Hydrocephaly | 741.90 | 741.91 | 741.92 | | |
| | | 741.93 | 741.94 | 741.98 | | |
| | | 741.985 | 741.99 | | | |
| A13 | Encephalocele | 742.00 | 742.08 | 742.09 | | |
| | | 742.085 | 742.086 | | | |
| A15 | Hydrocephaly | 742.30 | 742.31 | 742.38 | | |
| | | 742.39 | | | | |
| A16 | Microcephaly | 742.10 | | | | |

EYE AND EAR

| B03 | Glaucoma | 743.20 | 743.21 | 743.22 |
|-----------------|--|--------|---------|---------|
| B04 | Cataract | 743.32 | 743.325 | 743.326 |
| B51* | Anophthalmia | 743.00 | | |
| B52* | Microphthalmia | 743.10 | | |
| B54* | Ear anomaly with hearing loss | 744.00 | 744.01 | 744.02 |
| | | 744.03 | 744.09 | 744.21 |
| CARDIAC | | | | |
| D01 | Truncus Arteriosus | 745.00 | 745.01 | |
| D02 | Transposition of great vessels | 745.10 | 745.11 | 745.12 |
| | | 745.18 | 745.19 | |
| D03 | Tetralogy of Fallot | 745.20 | 745.21 | 746.84 |
| D04 | Single ventricle | 745.30 | | |
| D51* | Aortic stenosis | 746.30 | 746.31 | |
| D52* | Hypoplastic left heart | 746.70 | | |
| D53* | Total anomalous pulmonary venous | 747.42 | | |
| <u>RESPIRAT</u> | <u>CORY</u> | | | |
| E01 | Choanal atresia | 748.00 | | |
| E06 | Agenesis of lung | 748.50 | 748.51 | |
| <u>OROFACI</u> | <u>AL - GASTRO-INTESTINAL</u> | | | |
| F01 | Cleft palate | 749.00 | 749.01 | 749.02 |
| | | 749.03 | 749.04 | 749.05 |
| | | 749.06 | 749.07 | 749.09 |
| F02 | Cleft lip with or without cleft palate | 749.10 | 749.11 | 749.12 |
| | | 749.19 | 749.20 | 749.21 |
| | | 749.22 | 749.29 | |
| F08 | Pyloric Stenosis | 750.51 | | |
| | | | | |

| F09 | Tracheo-esophageal fistula or | 750.30 | 750.31 | 750.32 |
|-----------------|---------------------------------------|---------|---------|---------|
| | | 750.325 | 750.33 | |
| F14 | Stenosis or atresia of duodenum | 751.10 | | |
| F15 | Other stenosis or atresia of small | 751.11 | 751.12 | 751.19 |
| F16 | Stenosis or atresia of rectum or anus | 751.21 | 751.22 | 751.23 |
| | | 751.24 | | |
| F17 | Hirschsprung's Disease | 751.30 | 751.31 | 751.32 |
| | | 751.33 | | |
| F18 | Malrotation of intestine | 751.40 | 751.41 | 751.42 |
| | | 751.49 | 751.495 | |
| F21 | Biliary atresia | 751.65 | | |
| <u>GENITO-U</u> | JRINARY | | | |
| H01 | Renal agenesis | 753.00 | 753.01 | |
| H06 | Obstruction of kidney or ureter | 753.20 | 753.21 | 753.22 |
| | | 753.29 | 753.40 | 753.42 |
| H09 | Bladder or urethra obstruction | 753.600 | 753.61 | 753.62 |
| | | 753.63 | | |
| MUSCUL | <u>OSKELETAL</u> | | | |
| J03 | Dislocation of hip | 754.30 | | |
| J51* | Complete absence of upper or lower | 755.20 | 755.30 | 755.40 |
| J52* | Phocomelia of Limb | 755.21 | 755.31 | 755.41 |
| K05 | Amniotic bands | 658.80 | | |
| N01 | Diaphragmatic hernia | 756.61 | 756.615 | 756.616 |
| | | 756.617 | | |
| N02 | Omphalocele | 756.70 | | |
| N04 | Gastroschisis | 756.71 | | |
| | | | | |

SYNDROMES

| R01 | Down Syndrome (Trisomy 21) | 758.00 | 758.01 | 758.02 |
|-----|-------------------------------|--------|---------|---------|
| | | 758.03 | 758.04 | 758.09 |
| R02 | Patau Syndrome (Trisomy 13) | 758.10 | 758.11 | 758.12 |
| | | 758.13 | 758.19 | |
| R03 | Edwards Syndrome (Trisomy 18) | 758.20 | 758.21 | 758.22 |
| | | 758.23 | 758.29 | 758.295 |
| S02 | Fetal Alcohol Syndrome | 760.71 | 760.718 | |

* Codes created by CBDMP

| <u>PREC</u> | CISION (of diagnosis) (B | ox 32 | FORM 01) | | |
|-------------|---|-----------------|--|--|--|
| Code | | | | | |
| 1 | Not stated (For Mental Retardation and Cerebral Palsy Diagnoses ONLY - Form 03) | | | | |
| 2 | Probably not a birth defect ("Ruled out" included in this category), "NO" | | | | |
| 3 | "vs" (versus) or "or" | | | | |
| 4 | "Rule out" included in diagnos "Questionable," "R/O" | sis (i.e., rule | out anencephaly), "Doubtful," "Equivocal", | | |
| 5 | "Suggestive of" | | | | |
| б | "Suspected," "suspicious" | | | | |
| 7 | "Possible," "may have," "could be," "felt to be," "Perhaps," "consider" | | | | |
| 8 | "Consistent with," "most likely" | | | | |
| 9 | "Compatible with," "like," "appear | 'S'' | | | |
| 10 | "Probable," "presume" | | | | |
| 11 | | | | | |
| 12 | Precise diagnosis, "characteristic or | f' | | | |

13 Precise diagnosis with congestive heart failure or medicated with Digoxin, Drisdol, Chlorothiazide, Lasix, Lanoxin, Aldactone or diuretics (only for VSD, PDA, ASD, or Patent Foramen Ovale)

Abbreviations

| ABDMP | - Arizona Birth Defects Monitoring Program |
|-------|---|
| ADHS | - Arizona Department of Health Services |
| BPA | - British Pediatric Association |
| CBDMP | - California Birth Defects Monitoring Program |
| CDC | - Centers for Disease Control and Prevention |
| CRS | - Children's Rehabilitative Services (ADHS) |
| ICD | - International Classification of Disease |
| MACDP | - Metropolitan Atlanta Congenital Defects Program |
| | |

Exclusion List - ABDMP Non-reportable Birth Defects Cases

The following potential cases are not included in the ABDMP report for 1995:

- ! 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 referring to CDC and CBDMP list of `excludables.
- Babies born to mothers whose residence is out-of-state or out-of-country (i.e., nonresident cases).
- ! "Negatives," that is of cases ruled-out during case finding and medical record review.
- ! "No Match" cases: Birth Certificate was not on file and state of birth cannot 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 is no reliable denominator that can be used to generate a birth defect rate.
- Prenatally diagnosed cases that have not resulted in a 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 3 for list of Precision of Diagnosis codes.
- ! Cases only diagnosed outside of the hospital setting are not included in the ABDMP.

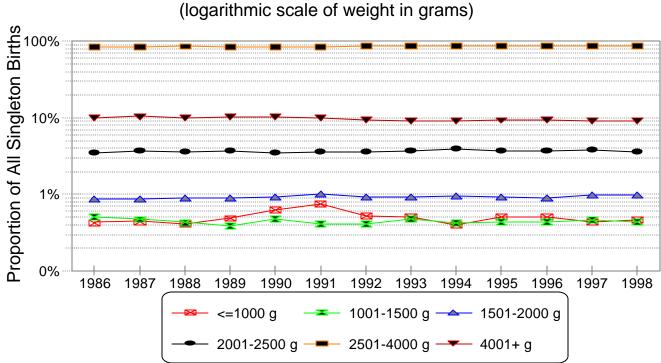
References

- 1 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.
- 2 Hexter AC, Harris JA, Roeper P, Croen LA, Krueger P, Gant D. Evaluation of the hospital discharge diagnoses index and the birth certificate as sources of information on birth defects. *Public Health Reports*. 1990; 105:296-306.
- Hexter AC, Harris JA. Bias in Congenital Malformations from the Birth Certificate. *Teratology*. 1991; 44:177-180.
- Mathis MP, Lavoie M, Hadley C. Birth Certificates as a Source for Fetal alcohol Syndrome Case
 Ascertainment Georgia, 1989-1992. *Morbidity and Mortality Weekly Report*. 1995; (44)13:251-253.
- 5 Calle EE, Khoury MJ. Completeness of the Discharge Diagnoses as a Measure of Birth Defects Recorded in the Hospital Birth Record. *American Journal of Epidemiology*. 1991; (134) 1:69-77.
- 6 Snell LM, Little BB, Knoll KA, Johnston WL, Rosenfeld CR. Reliability of Birth Certificate Reporting of Congenital Anomalies. *American Journal of Perinatology*. 1992; (9) 3:219-222.
- 7 Olsen CL, Polan AK, Cross PK. Case Ascertainment for state-based birth defects registries of unreported infants ascertained through birth certificates and their impact on registry statistics in New York State. *Pediatric and Perinatology Epidemiology*. 1996; 10:161-174.
- 8 Martin JA, .Smith BL, Matthews TJ, Ventura SJ. Births and Deaths: Preliminary Data for 1998. *National Vital Statistics Report.* October 25, 1999;(47)25.

- 9 Yoon PW, Olney RS, Khoury MJ, Sappenfield WM, Chavez GF, Taylor D. Contribution of Birth Defects and Genetic Diseases to Pediatric Hospitalizations. *Arch Pediatr Adolesc Med.* 1997;151:1096-1103.
- 10 Waitzman NJ, Romano PS, Scheffler RM, Harris JA. Economic costs of birth defects and cerebral palsy United States, 1992. *Morbidity and Mortality Weekly Report*. 1995;. 44; No. 37:695-699.
- 11 Harris JA, James, L. State-by-State Cost of Birth Defects-1992. *Teratology*. 1997; 56:11-16.
- 12 Edmonds LD. Birth Defect Surveillance at the State and Local Level. *Teratology*. 1997; 56:5-7.
- 13 Stierman L. Birth Defects in Eleven California Counties: 1990-1992. California Birth Defects Monitoring Program 1996.
- 14 Ibid.
- 15 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.
- 16 American Academy of Pediatrics. Folic Acid for the Prevention of Neural Tube Defects. *Pediatrics*. 1999;104:325-327.
- 17 Berry RJ, LiZ, Erickson D, LiS, Moore CA, Wang H, et al. Prevention of Neural-Tube Defects with Folic Acid in China. *The New England Journal of Medicine*. 1999;(341)20:1485-1490.
- 18 Mrela CK. Arizona Health Status ans Vital Statistics 1996. Arizona Department of Health Services, December, 1997.
- 19 Mrela CK. Arizona Health Status ans Vital Statistics 1998. Arizona Department of Health Services, December, 1999.

Birth Weight

The Arizona Birth Defects Monitoring Program monitors the distribution of birth weight. The data is obtainable from the birth certificate and may allow the detection of major shifts over time in the proportion of newborns with low birth weight. ^{18,19}



Birth Weight of Arizona Singletons

Other Defects Collected by the ABDMP Incidence Rates Per 1,000 Live Births and Fetal Deaths Arizona, 1995

| CODE | DEFECT GROUP | TOTAL | RATE |
|---|--|---|--|
| A00 A17 | CENTRAL NERVOUS SYSTEM Reduction Deformities of Brain | 61 | 0.84 |
| B00 B05 B06 B07 | EYE AND EAR Coloboma of Iris Absence of Iris Other Anomalies of Iris | 9 2 4 | 0.12 0.02 0.05 |
| D00 D05 D06 D07 D11 D12 D13 D18 D26 D27 D28 D29 D23 | CARDIAC Ventricular Septal Defect Ostium Secundum Type Atrial Septal Defect Endocardial Cushion Defect All Atrial Septal Defect Anomalies of Pulmonary Valve Tricuspid Atresia & Stenosis Congenital Mitral Stenosis Coarctation of Aorta Other Anomalies of Aorta Anomalies of Great Veins Eisenmenger's Syndrome Pulmonary Artery Atresia with Septal Defect | 141 84 33 69 17 10 40 51 29 2 1 | 1.93 1.15 0.46 0.95 0.23 0.14 0.55 0.70 0.40 0.03 0.01 |
| G00 G02 G03 | GENITAL ORGANS Hypospadias Epispadias | 179 3 | 2.46 0.04 |
| H00 H08 | URINARY BLADDER Exstrophy of the Urinary Bladder | 3 | 0.04 |
| J00 J10 | TOTAL MUSCULOSKELETAL DEFECTS Deformities of the Feet | 1 | 0.01 |
| K00 K01 K02 | ALL LIMB REDUCTIONS Absence/deformity of Upper Limb Absence/deformity of Lower Limb | 41 15 | 0.56 0.25 |
| L00 L03 | Anomalies of Spine | 62 | 0.85 |
| XOO | MISCELLANEOUS DEFECTS | 76 | 1.04 |

The data show that among the other defects collected by the ABDMP, the most frequent defect are hypospadias and ventricular septal defect.