

# Family Health Dataline

## IN THIS ISSUE:

- ✦ For the first time, the occurrence of major congenital anomalies is reported for Alaska.
- ✦ The most common category of anomaly in Alaska during 1996 was congenital heart defects, followed by genitourinary and gastrointestinal system anomalies.
- ✦ Alaska has higher than expected rates of Hirschsprung's Disease, ventricular septal defect, and pulmonary artery and valve anomalies.
- ✦ Alaska has lower than expected rates of tricuspid valve atresia and stenosis, hypoplastic left heart syndrome, and diaphragmatic hernia.

## Birth Defects in Alaska

The Epidemiology and Evaluation Unit of the Section of Maternal, Child and Family Health (MCFH) analyzed data on birth defects reported to the Alaska Birth Defects Registry. The Birth Defects Registry was established by Alaska State statute and reporting began during January 1996. The purpose of the present initial analysis is to establish baseline rates of 43 major congenital anomalies for Alaska using data on children born during 1996 and reported to the Alaska Birth Defects Registry; to compare these baseline rates to rates for other states; and to evaluate the birth defects rate among specific geographic regions and racial/ethnic groups in Alaska.

### *Methods*

The Alaska Birth Defects Registry is a passive surveillance system. Hospitals, physicians, early intervention programs, pediatric clinics and other health care providers serving children from birth through the age of six years are required to report contacts involving the diagnosis of a congenital anomaly. Reportable conditions include International Classification of Diseases, 9<sup>th</sup> revision (ICD-9) codes 740-760, plus metabolic disorders (such as phenylketonuria), infantile cerebral palsy, muscular dystrophy, various anemias, congenital hypothyroidism and neurofibromatosis. The 43 major anomalies discussed here (Table 1) are those identified by the National Birth Defects Prevention Network for all states to begin reporting consistently. This consistency enables comparisons between states when states use the same ICD-9 codes and reporting systems to identify anomalies.

Rates for Alaska were established using reports of congenital anomalies among children born during 1996 to mothers who were state residents at the time of delivery. Data presented here include only children born during 1996 and diagnosed with at least one of the listed congenital anomalies in the first year of life. The total number of live births to women who were Alaskan residents during 1996 ( $N = 10,041$ ) was obtained from the Alaska Bureau of Vital Statistics (BVS). Data used in the analyses were obtained from hospitals (reporting hospitals accounted for 90 percent of births statewide for 1996), Medicaid records, physicians

and pediatric clinics located in Anchorage and Fairbanks, state genetics clinics, specialty clinics and early intervention programs. Data on fetal deaths, stillbirths, or terminations are not included in these analyses.

Comparison rates were provided by tables published by the National Birth Defects Prevention Network (Edmonds, 1997). These tables provide the actual number of events of each of the anomalies reported, as well as the total number of births and descriptions of each states' surveillance methodology, allowing for calculation of relative risks for birth defects rates in Alaska compared to other states. Alaskan rates were compared to Colorado (using data reported for 1993) when Colorado collected data on the anomaly to be compared. When data from Colorado were not available, Alaskan data were compared to New Jersey (using data for 1992). All rates are reported per 1,000 live births. Colorado employs a passive surveillance system that includes only data on live births, similar to the system of passive surveillance in Alaska. New Jersey also employs a passive surveillance system that includes data on live births only.

We also compared the risk of having a specific birth defect among different racial and ethnic groups. Race/ethnicity was reported by providers as the race for the child at the time of diagnosis. In those cases where race/ethnicity was not collected or reported by the provider who made the diagnosis, race/ethnicity

**Table 1. Cases of major congenital anomalies per 1,000 live births, Alaska Birth Defects Registry, 1996.**

<i>Defect (ICD-9* code)</i>	<i>Cases per 1,000 live births (95% confidence interval)</i>
<b>Cardiovascular defects</b>	
Patent ductus arteriosus (747.0)	5.9 (4.5, 7.6)
Ventricular septal defect (745.4)	5.6 (4.2, 7.2)
Atrial septal defect (745.5)	4.5 (3.3, 6.0)
Pulmonary valve anomalies (747.3)	2.4 (1.5, 3.6)
Pulmonary valve atresia and stenosis (746.01, 746.02)	1.1 (0.54, 2.0)
Coarctation of aorta (747.10)	0.39 (0.11, 1.0)
Endocardial cushion defect (745.60, 746.61, 745.69)	0.29 (0.060, 0.87)
Tetralogy of Fallot (745.2)	0.19 (0.024, 0.72)
Common truncus (745.0)	0.19 (0.024, 0.72)
Aortic valve stenosis (746.3)	0.19 (0.024, 0.72)
Transposition of the great arteries (745.10, 745.11, 745.12, 745.19)	0.10 (0.025, 0.55)
Tricuspid valve atresia and stenosis (746.1)	0 (0, 0.30)
Ebstein's anomaly (746.2)	0 (0, 0.30)
Hypoplastic left heart syndrome (746.7)	0 (0, 0.30)
<b>Genitourinary system</b>	
Hypospadias and epispadias (752.61, 752.62)	3.1 (2.1, 4.4)
Obstructive genitourinary defect (753.2, 753.6)	1.4 (0.76, 2.3)
Renal agenesis/hypoplasia (753.0)	0.19 (0.024, 0.72)
Bladder exstrophy (753.5)	0 (0, 0.30)
<b>Gastrointestinal system</b>	
Pyloric stenosis (750.5)	2.2 (1.4, 3.3)
Hirschsprung's disease (751.3)	1.3 (0.69, 2.2)
Rectal and large intestinal atresia/stenosis (751.2)	0.69 (0.28, 1.4)
Cleft palate without cleft lip (749.00-749.04)	0.59 (0.22, 1.3)
Cleft lip with and without cleft palate (749.0, 749.2)	0.59 (0.22, 1.3)
Gastroschisis (756.79)	0.39 (0.11, 1.0)
Choanal atresia (748.0)	0.10 (0.025, 0.55)
Esophageal atresia/tracheoesophageal fistula (750.3)	0.10 (0.025, 0.55)
Biliary atresia (751.61)	0.10 (0.025, 0.55)
Diaphragmatic hernia (756.6)	0 (0, 0.30)

**Table 1. Continued**

<b>Defect (ICD-9* code)</b>	<b>Cases per 1,000 live births (95% confidence interval)</b>
<b>Central nervous system and cranium</b>	
Microcephalus (742.1)	1.1 (0.54, 2.0)
Hydrocephalus without spina bifida (742.3, w/o 741.0, 741.9)	0.29 (0.060, 0.87)
Anencephalus (740.0-740.1)	0.19 (0.024, 0.72)
Congenital cataract (743.30-743.34)	0.19 (0.024, 0.72)
Anotia/microtia (744.01, 744.23)	0.19 (0.024, 0.72)
Spina bifida without anencephalus (741.0, 741.9 w/o 740.0-740.10)	0.10 (0.025, 0.55)
Encephalocele (742.0)	0.10 (0.025, 0.55)
Anophthalmia/microphthalmia (743.0, 743.1)	0.10 (0.025, 0.55)
Aniridia (743.45)	0 (0, 0.30)
<b>Chromosomal</b>	
Down syndrome (758.0)	1.3 (0.69, 2.2)
Trisomy 18 (758.2)	0.10 (0.025, 0.55)
Trisomy 13 (758.1)	0 (0, 0.30)
<b>Other</b>	
Reduction deformity, lower limbs (755.30-755.39)	0.19 (0.024, 0.72)
Reduction deformity, upper limbs (755.20, 755.29)	0.10 (0.025, 0.55)
Lung agenesis/hypoplasia (748.5)	0.10 (0.025, 0.55)

*\*International Classification of Diseases, 9<sup>th</sup> Revision*

was obtained by linking to BVS data from birth certificates. Of the 303 unduplicated children with at least one major anomaly, race/ethnicity was available for 296 (missing/unknown = 7). Racial and ethnic groups evaluated included white, Alaska Native, black and other (including Hispanic, Asian, and Pacific Islander).

We also determined the risk of being born with a congenital anomaly by geographic area of the state. Community of residence at the time of diagnosis as reported by the health care provider was recoded into one of eight family health regions (Table 2). Relative risks for Down syndrome were calculated based on reports of maternal age at the time of the child's birth, which were obtained from BVS data.

## Results

A total of 1,214 unduplicated children born during 1996 (12 percent of total births) were reported to the Alaska Birth Defects

Registry. This is inclusive of all conditions, minor anomalies and various maternal conditions included in the 760 sequence, maternal infections, and prenatal exposure to cocaine and alcohol. Of these, there were 303 unduplicated children born with at least one of the 43 major anomalies (Table 1). The anomalies with the highest incidence rates were patent ductus arteriosus, ventricular septal defect, atrial septal defect and hypospadias and epispadias.

We identified four anomalies for which the number of cases per 1,000 live births was significantly higher than that found in comparison states (Table 3), including pulmonary artery anomaly and Hirschsprung's disease. Alaska had significantly more cases per 1,000 live births than either comparison state. Additionally, no children born in Alaska during 1996 had tricuspid valve atresia and stenosis, hypoplastic left heart syndrome, or diaphragmatic hernia, although data from other states suggest that Alaska should have had some cases.

Similar to other states, we found that Down syndrome occurred more commonly among mothers older than 34 years of age when compared to younger mothers (RR=4.18; 95% CI=1.37, 12.76).

Among racial/ethnic groups, major anomalies were more common among Alaska Natives and blacks (Table 4). Among Family Health Regions, only the Yukon-Kuskokwim Delta region showed a significantly different relative risk of any major

anomaly than Anchorage. The Yukon-Kuskokwim Region had fewer major anomalies per 1,000 live births than Anchorage (Table 5). When combined, 181 of 5,577 live births in Anchorage and Fairbanks had a major anomaly compared with 122 of 4,464 live births from other regions of the state (RR 1.19; 95% CI= .95, 1.49).

### ***Discussion and Limitations***

The Alaska Birth Defects Registry was designed with several goals in mind. These include identifying the number of children statewide who have congenital anomalies, describing the severity of the conditions that these children experience, identifying the children who are not accessing available services and identifying needs that are not being met through the services currently available. This presentation of initial results is the first step toward identifying the number and types of congenital anomalies present in children born in Alaska during 1996. Data collection and analysis will continue to determine the accuracy of the baseline occurrence of reported anomalies; additionally, we will validate results through active medical chart review. Once data reporting becomes routine for providers, and case ascertainment and quality control procedures are firmly in place, the next action will be to determine which children reported by medical providers are enrolled in medical service delivery systems such as the State Genetics Clinics and the Early Intervention Programs. This will allow us to identify

**Table 2. Family Health Regions as specific collections of census areas.**

<i>Region</i>	<i>Census Areas</i>
Anchorage	Anchorage
Fairbanks North Star Interior	Fairbanks North Star Borough Denali Borough, SE Fairbanks, Yukon-Koyukuk
North/Northwest	Nome, North Slope, Northwest Arctic Borough
South Central	Kenai Peninsula, Matanuska Susitna, Valdez-Cordova
Southeast	Haines, Juneau, Ketchikan Gateway, Prince of Wales, Sitka, Skagway/Hoonah/Angoon, Yakutat, Wrangell-Petersburg
Southwest	Aleutians East, Aleutians West, Bristol Bay, Dillingham, Kodiak Island, Lake and Peninsula
Yukon-Kuskokwim	Bethel, Wade Hampton

gaps in service delivery and subsequently to improve access to services for children with congenital anomalies.

Problems of case ascertainment are inherent in passive surveillance systems, particularly with regard to their sensitivity. Sensitivity is defined as the probability of being identified with a birth defect when a birth defect is present. As noted in Table 1, the lack of any cases reported for some of the major anomalies may be due to use of an out-of-state provider. It is possible that infants with serious defects were transported to facilities in Washington or Oregon either immediately before or after birth. These cases would not be detected using the current surveillance system, as out-of-state providers are not reporting contacts. If the child did not return to Alaska, (e.g., if he or she died due to a lethal anomaly), the child would not be picked up in reports by an in-state provider. Lack of sensitivity could also be due to specific diagnostic practices in Alaska (for example, if postnatal ultrasound were used less in Alaska than in other states, it might lead to fewer diagnoses of ventricular septal defects) and failure to include fetal deaths in our surveillance system. To address these issues, we have begun including data from fetal and infant deaths and coordinating with out-of-state providers to increase the sensitivity of our surveillance system.

There are also problems inherent in comparing birth defects rates between states, even if they use the same surveillance system design. For example, physician diagnostic patterns may differ or migration of different sub-populations between states may occur due to employ-

**Table 3. Relative risks for selected major congenital anomalies in Alaska compared to either Colorado\* or New Jersey,\*\* Alaska Birth Defects Registry, 1996 births.**

<i>Defect</i>	<i>Cases per 1,000 live births</i>			<i>Relative risk (95% confidence interval)</i>
	<i>Alaska</i>	<i>Colorado</i>	<i>New Jersey</i>	
Ventricular septal defect (745.4)	5.6	3.0		1.9 (1.4, 2.6)
Pulmonary valve atresia & stenosis (746.01, 746.02)	1.1		0.56	2.0 (1.0, 3.7)
Pulmonary artery anomalies (747.3)	2.4	0.76		3.2 (1.9, 5.2)
Hirschsprung's disease (751.3)	1.3	0.20		6.4 (2.9, 14)

\*1993 data \*\*1992 data

ment patterns or other factors. Nevertheless, these data provide a reference point useful for interpreting Alaska rates.

In particular, we found unexpectedly high rates of some major anomalies in Alaska. For these anomalies, we will conduct active case ascertainment. Review of medical charts for Hirschsprung's disease cases is scheduled to begin during December 1998. While the etiology of the condition is not well understood, previous research has shown that cases of Hirschsprung's disease tend to run in families, tend to occur in children whose mothers are younger than 35 years of age, and are significantly less likely to occur in first born children (Torfs, 1996; Badner et al., 1990; Ryan et al., 1992). Further investigation of Alaskan cases is

necessary to determine if these or other associations are discernible for Alaska.

Differences noted between geographic regions within Alaska should be viewed with caution at this early stage of the Registry. Reporting on birth defects is new, and differences in the way data are recorded exist between regions of the state (for example, computerized versus non-computerized). This is consistent with our finding that reported rates of major anomalies were considerably lower outside of the two major urban regions of the state. In some parts of the state, we expect a "learning curve" effect, as providers become more familiar with reporting requirements. Additionally, for rural areas of the state not in close proximity to Anchorage or Fairbanks, we are relying almost exclusively on data provided by hospitals. As other providers in those areas begin reporting, we can expect to see an increase in the number

**Table 4. Relative risks for any major congenital anomaly by race/ethnicity, Alaska Birth Defects Registry, 1996 births.**

<i>Race/Ethnicity</i>	<i>Relative risk</i>	<i>95% Confidence interval</i>
White	Ref.	-
Alaska Native	1.4	1.1, 1.8
Black	1.9	1.3, 3.1
Other	0.85	0.47, 1.6

of cases identified.

Before the establishment of the Alaska Birth Defects Registry, few data existed on the incidence of birth defects in Alaska. The establishment of the Registry and its development as a sensitive and representative measurement of the burden of birth defects in Alaska will provide the basis for further understanding of the occurrence of birth defects, identification of high risk groups, and service provision to affected children. Work is currently underway to determine rates among children born in 1997.

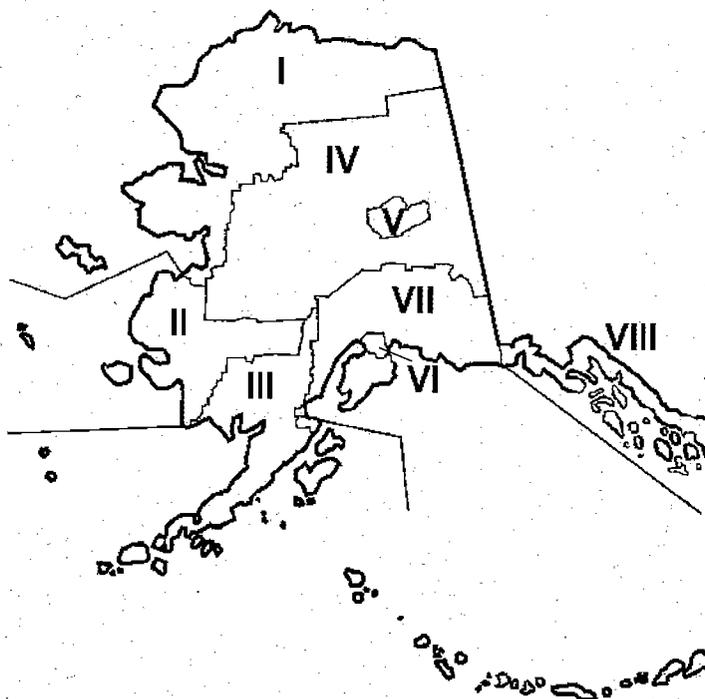
*Contributed by  
Grace Reynolds, MPA  
and Brad Gessner, MD, MPH*

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**Table 5. Relative risks for any major congenital anomaly by family health region, Alaska Birth Defects Registry 1996.**

<i>Geographic area</i>	<i>% of total births in area reported with at least one major anomaly</i>	<i>Relative risk (95% confidence interval)</i>
Anchorage (VI)	3.2	Ref.
Fairbanks (V)	3.1	0.96 (0.69, 1.30)
Interior (IV)	5.5	1.60 (0.91, 2.90)
Northwest (I)	2.7	0.84 (0.49, 1.40)
South Central (VII)	2.7	0.84 (0.60, 1.20)
Southeast (VIII)	2.7	0.83 (0.56, 1.20)
Southwest (III)	2.6	0.80 (0.45, 1.40)
Yukon-Kuskokwim (II)	1.8	0.53 (0.29, 0.98)



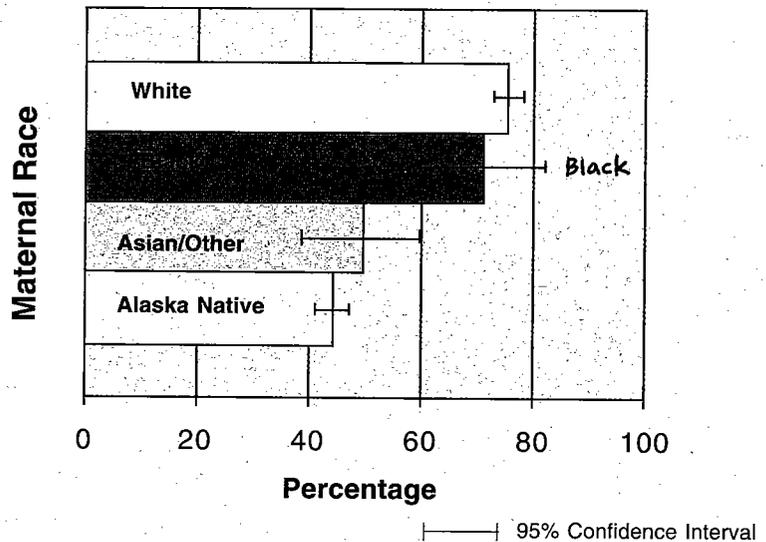
# Folic Acid Knowledge in Alaska

Neural tube defects (NTDs) occur in approximately one out of every 1,000 births in the United States. NTDs are severe birth defects, involving incomplete development of the brain, spinal cord or the protective coverings of these organs. The use of folic acid supplementation in women of childbearing age has been well documented as a means of reducing NTDs.<sup>1,2</sup> The United States Public Health Service issued recommendations that every woman capable of becoming pregnant should consume 400 micrograms of folic acid per day.<sup>3</sup>

Through the Pregnancy Risk Assessment Monitoring System (PRAMS), a population-based survey of mothers of newborns, MCFH has data on the extent to which Alaskan women are knowledgeable concerning folic acid's role in reducing birth defects.

PRAMS was developed by the Centers for Disease Control and Prevention (CDC) to gather information on the health risk behaviors and circumstances of pregnant and postpartum women. A systematic, stratified sampling approach is used to select approximately one of every six mothers of newborns each month from the state's live birth records. During 1996 and 1997 birth years, 2,422 women responded to the PRAMS survey – a 75% response rate. The percentages given reflect statewide

**Figure 1. Folic acid knowledge among mothers of newborns by race, Alaska PRAMS, 1996-1997**



estimates for 18,018 Alaska-resident women who delivered a live birth during 1996 or 1997.

The current PRAMS questionnaire elicits information on folic acid knowledge by asking, "Have you heard or read that taking the vitamin folic acid can help prevent some birth defects?" The overall percent of women knowledgeable about folic acid for the two-year period was 67%. Data for 1996 show that 63% of women were knowledgeable about folic acid's ability to prevent some birth defects. Data for 1997 showed that 70% of women had knowledge of folic acid's benefits, a statistically significant increase.

Knowledge of folic acid's beneficial effects was higher for white and black mothers (76% and 71%, respectively) than for Alaska Natives and Asians or another race (44% and 50%, respectively) (Figure 1).

The PRAMS data will provide a temporal measure of folic acid knowledge among Alaska-resident women who have delivered a live birth. It is especially advantageous that the first year of data which PRAMS provides on folic acid knowledge (1996) is a true baseline, having been collected for the same initial birth cohort as the Birth Defects Registry and prior to extensive intervention efforts. Health care providers who serve women of child-bearing age should continue to share information on the benefits of folic acid supplementation. For materials please contact the Birth Defects Registry Coordinator (see page 8).

*Contributed by Kathy Perham-Hester, MS, MPH and Grace Reynolds, MPA*

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To contact the Alaska Birth Defects Registry or obtain information on folic acid supplementation, call or write:

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