Hawai‘i State Department of Health
Hawai‘i Birth Defects Program

Hawai‘i Birth Defects Surveillance Report

1986-2005

Aloha!

Birth defects can have a significant impact on children and their families. Major structural and genetic birth defects may result in serious illness, developmental delays, long term disability, and/or death. As a result, more health care, education, and social services may be needed throughout childhood and adolescence and into adult life.

The Hawaiʻi State Department of Health is pleased to present 20 years of data in this Hawaiʻi Birth Defects Surveillance Report for 1986-2005. The report highlights trends in birth defects and variation by county of residence, maternal ethnicity, prenatal care, and other factors.

It is hoped that birth defects data may be useful for developing public health policy and community strategies to prevent birth defects. Data may also be used in planning services to ensure that children with birth defects have the necessary services for optimal health, growth, and development.

Attention to a vulnerable population of children with special health care needs contributes toward improving the health of all children in Hawaiʻi.

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BIRTH DEFECTS DATA IN HAWAI‘I, 1986-2005
Summary

Annual rate	4.4% of births have a major structural and/or genetic birth defect. The most common birth defect categories are cardiac & circulatory, limb & musculoskeletal, and genital & urinary conditions. Of birth defect cases, 30% involve two or more categories of birth defects.
Trends	Birth defect trends from 1986 to 2005 were varied, with 12 birth defect conditions having significant increasing trends, 44 having no change, and 41 having significant decreasing trends. Of note is that neural tube defects, which can be prevented by maternal use of folic acid, showed a significantly decreasing trend.
Gender	Birth defect rate for males was 36% higher than that for females.
Maternal ethnicity	Birth defect rates varied by maternal ethnicity. The highest rates were in Black, Portuguese, Samoan, and “Other” women, and the lowest rate was in Vietnamese women.
Maternal residence	Birth defect rates for Hawai‘i and Maui Counties were similar and significantly lower compared to the rates for Kauai County and the City and County of Honolulu. Birth defect rates for urban and rural areas were similar.
Gestational age	Birth defect rates for infants born early at 20-37 weeks gestational age were significantly higher than for infants born at ≥38 weeks.
Maternal age	Birth defect rates were similar for maternal age groups up to age 34 years, with higher rates for older mothers age ≥35 years.
Prenatal care	Birth defect rate for mothers who received no prenatal care was over 2.5 times higher than the rate for mothers who received some prenatal care.
Maternal substance use	Compared to mothers who were non-users: ▪ Birth defect rates were over two-fold higher for mothers who used tobacco. ▪ Birth defect rates were six-fold higher for mothers who used alcohol. ▪ The estimated proportion of birth defects was 52% higher for mothers who used illicit drugs.
Infant death	Of live births with birth defects, 4.9% were associated with infant death. The number of infant deaths associated with birth defects is 2.1 per 1,000 live births. Of all infant deaths, 29.6% were associated with birth defects.

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OVERVIEW OF BIRTH DEFECTS SURVEILLANCE IN HAWAIʻI

The Hawaiʻi Birth Defects Program (HBDP) provides population-based surveillance for birth defects in Hawaiʻi. HBDP monitors major structural and genetic birth defects that adversely affect health and development.

Birth defects can have a significant impact on children and their families. Birth defects may result in serious illness, developmental delays, long term disability, and/or death. There may be a long-term need for health, education, and social services. Medical costs can be high. Hospital costs for birth defects in the U.S. are over $2.5 billion per year.[1]

Birth defects surveillance data may assist in improving birth outcomes and the health of Hawaii's children. Data may be used for developing baseline rates, monitoring trends, and performing cluster investigations. Data may be used by public health and community programs in developing policies, developing prevention strategies, planning services, and program evaluation.

Hawaiʻi Birth Defects Program

HBDP was established in August 1988 in response to a community-based recommendation to the Governor to establish a reliable birth defects and adverse reproductive outcome surveillance system for the State of Hawaiʻi. HBDP is a program within the Children with Special Health Needs Branch, Family Health Services Division, Hawaiʻi State Department of Health (DOH).

HBDP responsibilities are established by law, as defined in Hawaiʻi Revised Statutes (HRS) §321.421-426 and HRS §324.41-44. HBDP is required to:

- Collect surveillance information of birth defects and other adverse reproductive outcomes;
- Report incidence, trends, and causes of birth defects and other adverse reproductive outcomes;
- Report information for the development of prevention strategies; and
- Develop strategies to improve access to health and early intervention services for children with birth defects.

HBDP conducts birth defects surveillance in its capacity as a Public Health Authority as defined by the Health Insurance Portability and Accountability Act (HIPAA) Privacy Rule. Security measures have been established to protect the privacy of individually identifiable information. Only de-identified data are used in surveillance reports.

The birth defects surveillance system in Hawaiʻi is an active surveillance system in which birth defects data are directly abstracted from medical and other records at hospitals and other medical facilities statewide.

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[1] Agency for Healthcare Research and Quality, Healthcare Cost and Utilization Project, Statistical Brief #24, "Hospitalizations for Birth Defects, 2004," January 2007. http://www.hcup-us.ahrq.gov/reports/statbriefs/sb24.pdf

Guidelines for Birth Defects Surveillance
HBDP follows the "Guidelines for Conducting Birth Defects Surveillance"[2] of the National Birth Defects Prevention Network (NBDPN), a national network of state and birth defects programs supported by the Centers for Disease Control and Prevention (CDC). NBDPN guidelines include the CDC classification of birth defects relating to the brain and nervous system; eye, ear, face, and neck; cardiac and circulatory; respiratory; orofacial and gastrointestinal; genital and urinary; limb and musculoskeletal; and skin and integument systems. The diagnostic codes used are the British Pediatric Association (BPA) system, which is similar to the International Classification of Diseases (ICD), but with more digits to specify a more precise diagnosis (Appendix A).

Case Definition
All infants and fetuses with birth defects are included regardless of pregnancy outcome (live birth, fetal demise, elective or spontaneous termination).

For an infant/fetus to be included in the birth defects registry, the following criteria are used:

1. The infant/fetus must have one or more reportable diagnoses. Reportable conditions include over 1,000 types of moderate to severe birth defects, as defined by the CDC, as well as neonatal tumors and fetal alcohol syndrome (FAS).
2. The end of the pregnancy must occur in the state of Hawai‘i, but conception and/or any part of the pregnancy may occur outside of the state.
3. The infant or fetus was noted to have a problem during the antenatal period or within one year after delivery (with the exception of FAS).
4. Cases of FAS are included in the birth defects registry without regard to the age of diagnosis. This is due to the public health interest in FAS. It should be noted that not all cases of FAS may be in the HBDP database, since FAS may be diagnosed after the first year of life and information may be in medical records that are not abstracted by HBDP.

Data Abstraction
Potential cases are identified through examination of discharge lists with certain conditions and other reports provided by hospitals in Hawai‘i where births typically occur, as well as at diagnostic clinics, cytogenetic laboratories, genetic counseling centers, and maternal-fetal centers in the state. Participating health care facilities are listed in Appendix B.

HBDP staff reviews both the infant's and mother's medical records to determine if the case meets birth defects case definition. If it does, the HBDP staff abstract data, which may include over 125 items of demographic and health related information on the infant/fetus and biological parents, including race/ethnicity, previous pregnancies affected by birth defects, and prenatal use of alcohol, tobacco, and illicit drugs (either by self-report, physician note, or positive toxicology screen). If the record indicates care at another Hawai‘i health care facility, then records at that facility are also abstracted to ensure completeness of the data. All data are reviewed by HBDP staff for completeness before transferring the information to the main database.

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[2] National Birth Defects Prevention Network (NBDPN). *Guidelines for Conducting Birth Defects Surveillance.* Sever LE, ed. Atlanta, GA: National Birth Defects Prevention Network, Inc., June 2004, http://www.nbdpn.org/birth_defects_surveillance_gui.php. The CDC listing of reportable birth defects is in Appendix 5.2.

To ensure that birth defect reporting is complete and accurate:

- Multiple data sources are used to obtain comprehensive information.
- Only cases with confirmed diagnoses are abstracted.
- Standardized codes for data collection variables are used for abstraction.
- Five percent (5%) of medical record charts are re-abstracted by HBDP field staff to determine completeness and accuracy.
- Ten percent (10%) of abstracts are reviewed for completeness and accuracy by program and/or medical personnel.

Data Interpretation

Case: A case is an individual infant/fetus with one or more birth defects. A case with more than one birth defect condition may be counted in two or more categories of birth defects.

Rate: A rate is used to adjust for differences in population sizes. For example, a rate of birth defects per 10,000 live births may be used to compare groups of different sizes.

Proportion: In a proportion, the cases in the numerator must be included in the denominator. A percentage is a proportion multiplied by 100.

Small Number: Rates and proportions based on fewer than 20 reported cases are not considered reliable due to small numbers and potential fluctuation from year to year. In particular, birth defect conditions with less than five reported cases need to be interpreted with caution.

Confidence Interval: Rates may be reported with confidence intervals. A 95% confidence interval indicates that the true rate should be contained in this interval 95% of the time. If rates are compared and their confidence intervals do not overlap, the difference between rates is considered meaningful or statistically significant. However, if the confidence intervals overlap, the difference between rates may be due to random fluctuation (or chance) and is probably not meaningful.

Trend: To determine trends for specific birth defects over time, trend lines were added to graphs of annual rates. The trend was modeled using a log-linear regression model (which is appropriate for data following a Poisson distribution). To determine statistical significance of a trend, the confidence interval for the weighted average of the first three years of rates were compared with the weighted average of the last three years of rates.

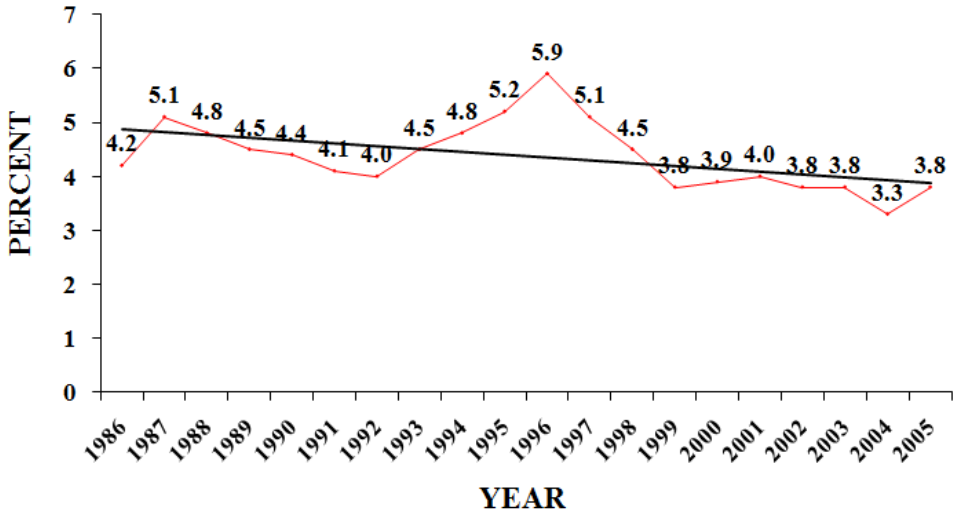
BIRTH DEFECTS DATA FOR HAWAI'I, 1986-2005

Proportion of Births with Birth Defects

For 1986-2005, the average annual proportion of births with birth defects is 4.4% (Table 1).

The birth defect proportion peaked 1995-1997 (5.9% in 1996). Although the overall trend shows a reduction in birth defects from an average of 4.7% for 1986-1988 to an average of 3.6% for 2002-2005, it is not statistically significant[3] (Figure 1).

FIGURE 1. PROPORTION OF TOTAL BIRTHS* WITH BIRTH DEFECTS, HAWAI'I, 1986-2005



*Total births = live births + fetal deaths

[3] Weighted average proportions were 4.7% (Confidence Interval [CI] 4.0-5.4) for 1986-1988 and 3.6% (CI 3.0-4.3) for 2003-2005.

TABLE 1. PROPORTION OF TOTAL BIRTHS WITH BIRTH DEFECTS, HAWAI'I 1986-2005

YEAR	TOTAL BIRTHS*	BIRTH DEFECT CASES**	
	Number (N)	Number (N)	Percent (%)***
1986	19,819	825	4.2
1987	20,097	1,027	5.1
1988	20,541	979	4.8
1989	20,954	943	4.5
1990	22,288	983	4.4
1991	21,765	903	4.1
1992	21,504	868	4.0
1993	21,073	932	4.4
1994	20,814	1,007	4.8
1995	19,750	1,024	5.2
1996	19,386	1,135	5.9
1997	18,198	925	5.1
1998	18,644	848	4.5
1999	18,067	693	3.8
2000	18,491	729	3.9
2001	17,868	722	4.0
2002	18,183	699	3.8
2003	19,033	717	3.8
2004	19,234	637	3.3
2005	18,875	715	3.8
Total	394,584	17,311	4.4

* Total births = live births + fetal deaths.

** Birth defect case = live birth, fetal death, or temination with one or more birth defect diagnoses.

*** Percent = (number of birth defect cases)/(total births)x100.

Birth Defect Rates

Of the broad categories for birth defects, the three most common categories are cardiac & circulatory (179.6 per 10,000 total births), limb & musculoskeletal (124.9), and genital & urinary conditions (103.8) (Figure 2A).

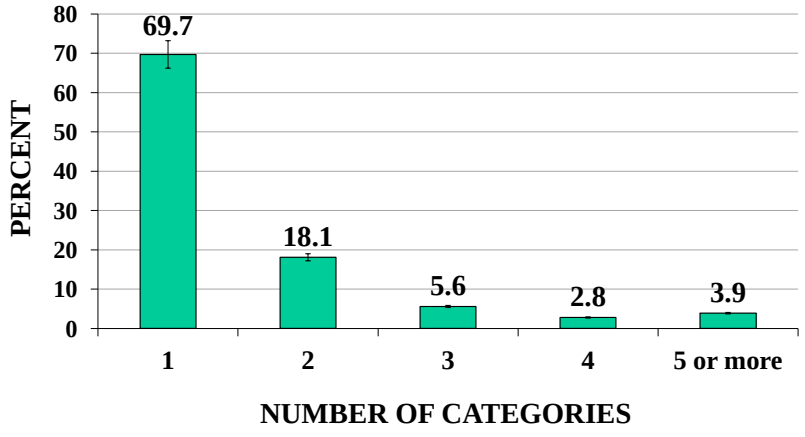
There are also differences in rates among specific birth defect conditions (Table 2, page 29).

FIGURE 2A. BIRTH DEFECT RATES PER 10,000 TOTAL BIRTHS[4] BY BROAD CATEGORIES[5], HAWAI'I, 1986-2005

Category	Rate
Cardiac & circulatory	179.6
Limb & musculoskeletal	124.9
Genital & urinary	103.8
Skin & integument	58.2
Orofacial & gastrointestinal	56.4
Chromosome & syndrome	52.9
Eye, ear, face, & neck	48.0
Brain & nervous system	39.1
Respiratory	23.8
Neoplasms	9.5

Of birth defect cases, 69.7% have birth defect(s) involving only one category of birth defects (Figure 2B). Of the remaining cases, 26.5% involve 2-4 categories, and 3.9% involve 5 or more categories.

FIGURE 2B. PROPORTION OF BIRTH DEFECT CASES INVOLVING MULTIPLE CATEGORIES OF BIRTH DEFECTS, HAWAI'I, 1986-2005



[4] Total births = live births + fetal deaths.
[5] A birth defects case with more than one diagnosis may be included in multiple categories.

From 1986 to 2005, trends in birth defect rates varied (Figure 2C and 2E). Of the birth defects listed:

- 12 birth defects had significant increasing trends.
- 44 birth defects had no change.
- 41 birth defects had significant decreasing trends.

Neural tube defects are among the birth defects showing a significantly decreasing trend. This decreasing trend may due to the increased use of folic acid which, if taken before conception and in early pregnancy, lowers the risk of having a baby with neural tube defects.[6]

The trend for fetal alcohol syndrome (FAS) significantly decreased. However, this may be due to underidentification and data collection issues, since FAS may be diagnosed after the first year of life and information may be in the medical records that are not abstracted by HBDP.

FIGURE 2C. TRENDS FOR BIRTH DEFECTS, HAWAI'I, 1986-2005[7]

Increasing Trend	No Change	Decreasing Trend
Brain and Nervous System		
	Spina bifida Encephalocele Holoprosencephaly	Neural tube defects Anencephaly Microcephaly Hydrocephaly (without spina bifida)
Eye		
	Anophthalmia Cataract	Microphthalmia
Ear		
Absent auditory canal/ ear	Microtia	
Cardiac & Circulatory		
Transposition of the great vessels Atrial septal defect Patent ductus arteriosus Pulmonary artery atresia/stenosis	Common truncus Tetralogy of Fallot Tricuspid valve atresia/ stenosis Ebsteins's anomaly Coarctation of aorta Aortic stenosis Total anomalous pulmonary venous return Partial anomalous pulmonary venous return	Single ventricle Ventricular septal defect Endocardial cushion defect Pulmonary valve atresia/stenosis Aortic valve atresia/ stenosis Mitral valve atresia/ stenosis Hypoplastic left heart syndrome Dextrocardia Persistent fetal circulation
Respiratory		
Lung cysts		Choanal atresia/ stenosis Hypoplastic lung
Orofacial & Gastrointestinal		
Atresia/stenosis small intestine	Cleft palate (isolated) Anal atresia/stenosis Biliary atresia	Oral clefts Cleft lip (± cleft palate) TE fistula/Esophageal atresia Pyloric stenosis Hirschprung's disease Malrotation
Genital & Urinary		
Hypospadias/ epispadias/chordee Hydronephrosis	Ovarian cyst Ambiguous genitalia Cystic kidney Exstrophy bladder Posterior urethral valves	Undescended testicle Absent/hypoplastic kidney

Continued

[6] http://www.cdc.gov/ncbddd/folicacid/index.html

[7] For significant increasing and decreasing trends, the weighted average rate for 1986-1988 was significantly different from the weighted average rate for 2003-2005 at the 95% confidence interval.

From 1986 to 2005, there were significant increasing trends for rates of birth defect cases with maternal use of marijuana, methamphetamine/amphetamine, and tobacco (Figure 2D and 2E). There were significant decreasing trends for rates of birth defect cases with maternal use of cocaine and alcohol.

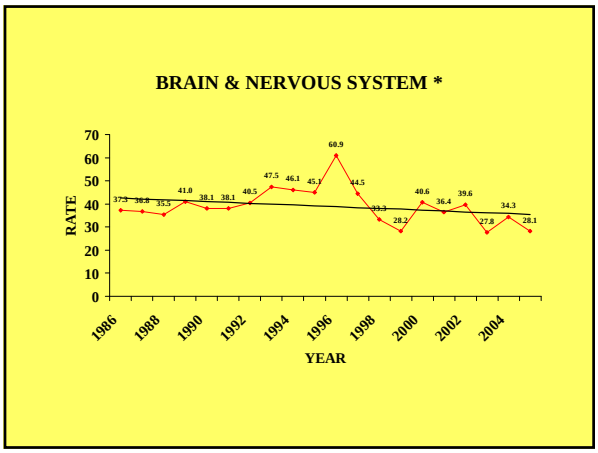
FIGURE 2C. TRENDS FOR BIRTH DEFECTS, HAWAI'I, 1986-2005 *(continued)*

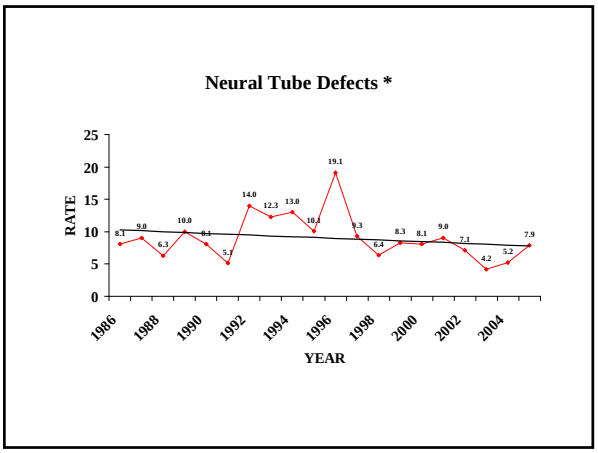
Increasing Trend	No Change	Decreasing Trend
Limb & Musculoskeletal		
Gastroschisis	Limb reduction deformity - lower Crouzon's disease Apert syndrome Goldenhar syndrome Spina bifida occulta Osteogenesis imperfecta Abdominal wall defects Omphalocele Prune-belly syndrome Poland syndrome	Scoliosis Hip dislocation Clubfoot Polydactyly Syndactyly Limb reduction deformity - upper Craniosynostosis Dwarfism Diaphragmatic hernia
Skin & Integument		
		Skin & integument
Chromosome & Syndrome		
Patau syndrome (trisomy 13) Werdnig Hoffman syndrome	Edwards syndrome (trisomy 18) Cri du Chat syndrome Wolf-Hirschorn syndrome Triploidy Turner syndrome (45,X) Kleinfelter syndrome (47,XXY) Situs inversus Conjoined twins Tuberous sclerosis DiGeorge syndrome Pierre-Robin sequence Wolff-Parkinson-White syndrome	Down syndrome (trisomy 21) Autosomal anomalies in normal infant Absent spleen/Aplasia Accessory spleen Amniotic band syndrome
Other		
		Neoplasms Fetal alcohol syndrome

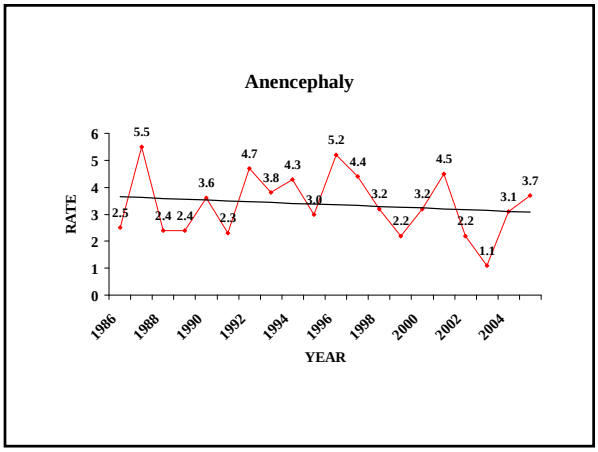
FIGURE 2D. TRENDS FOR BIRTH DEFECT CASES WITH MATERNAL SUBSTANCE USE, HAWAI'I, 1986-2005

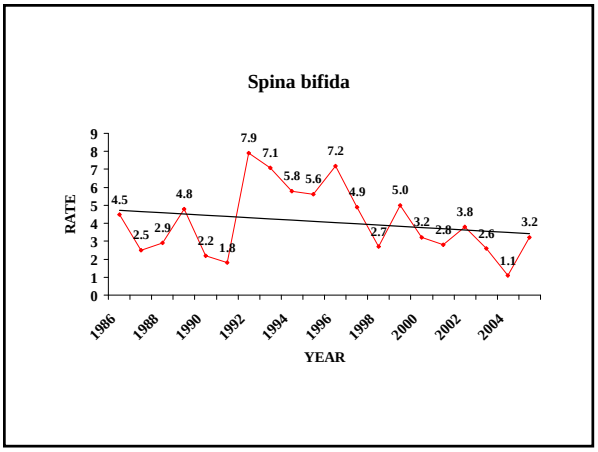
Increasing Trend	Decreasing Trend
Birth defects with maternal use of: Marijuana Methamphetamine/amphetamine Tobacco	Birth defects with maternal use of: Cocaine Alcohol

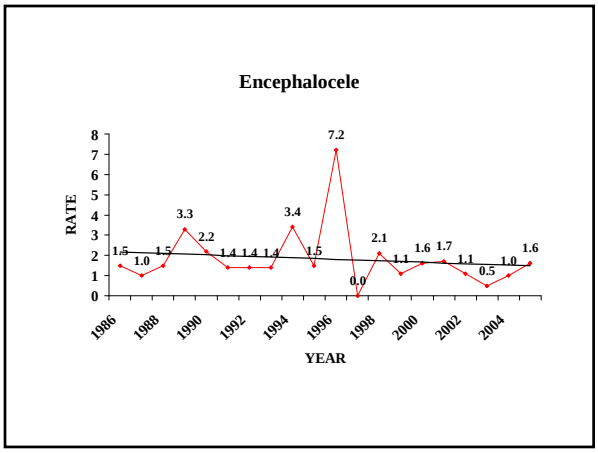
FIGURE 2E: TREND GRAPHS FOR BIRTH DEFECTS, HAWAI‘I, 1986-2005

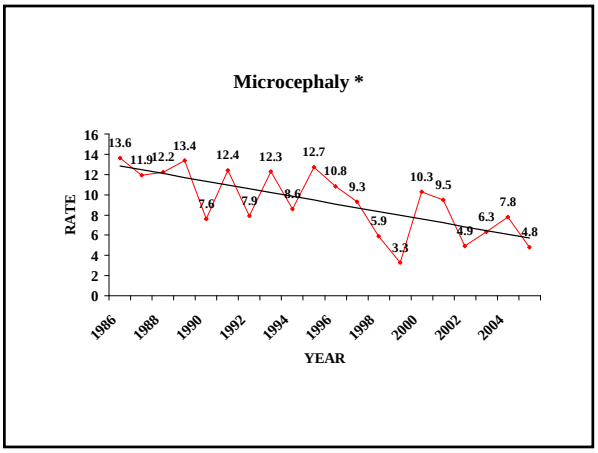






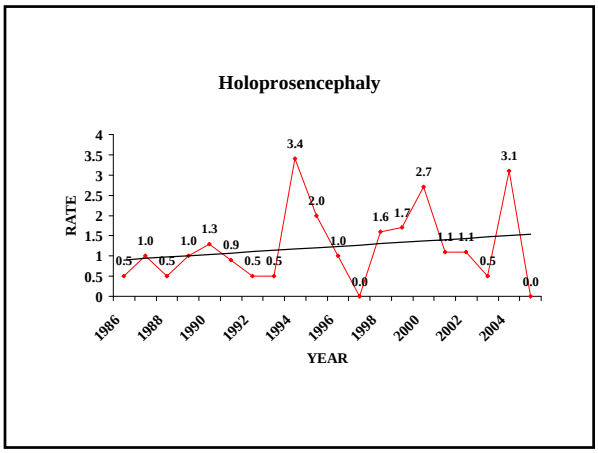


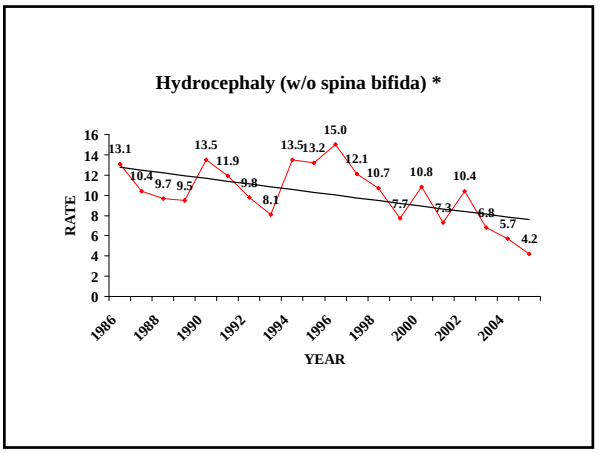


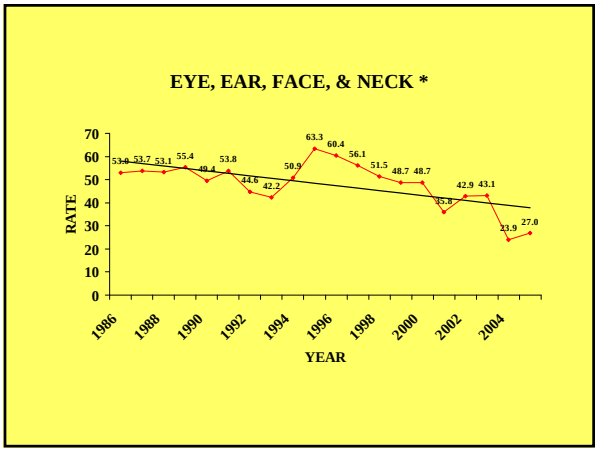


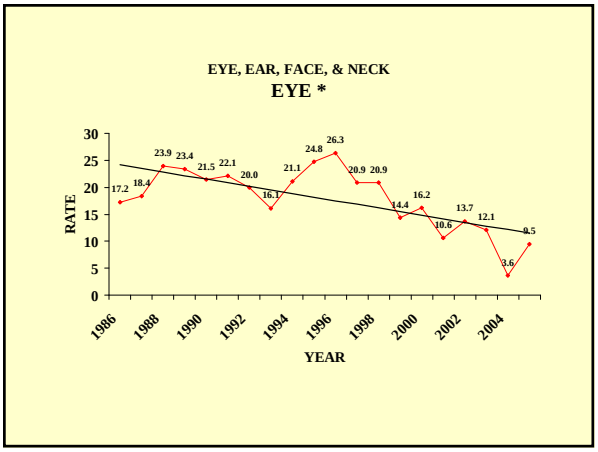
* *Weighted average rate for 1986-1988 was significantly different from weighted average rate for 2003-2005 at 95% C.I.*
Yellow background indicates categories for birth defects. Green background indicates categories for birth defect cases.

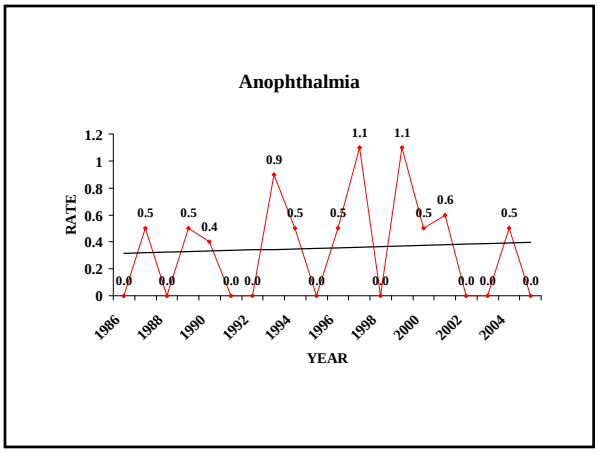
FIGURE 2E: TREND GRAPHS FOR BIRTH DEFECTS, HAWAI‘I, 1986-2005

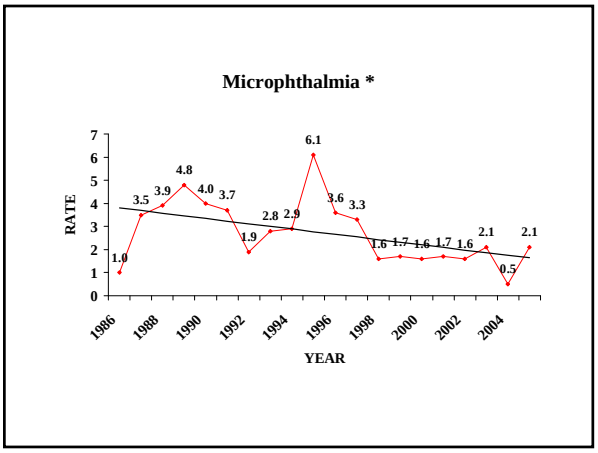












** Weighted average rate for 1986-1988 was significantly different from weighted average rate for 2003-2005 at 95% C.I.*
Yellow background indicates categories for birth defects. Green background indicates categories for birth defect cases.

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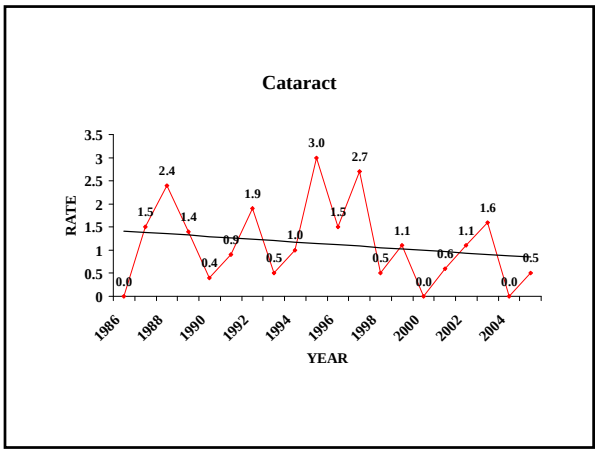

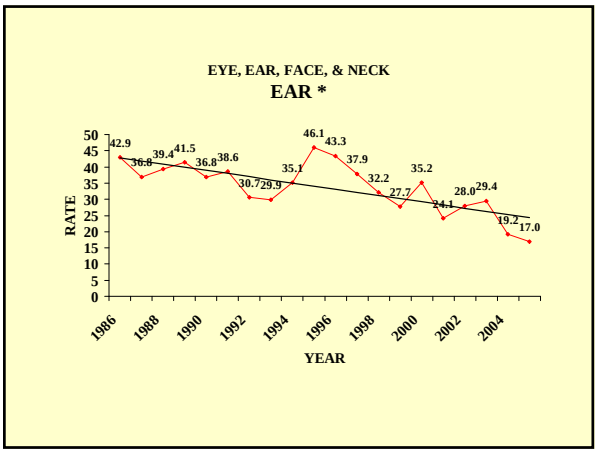

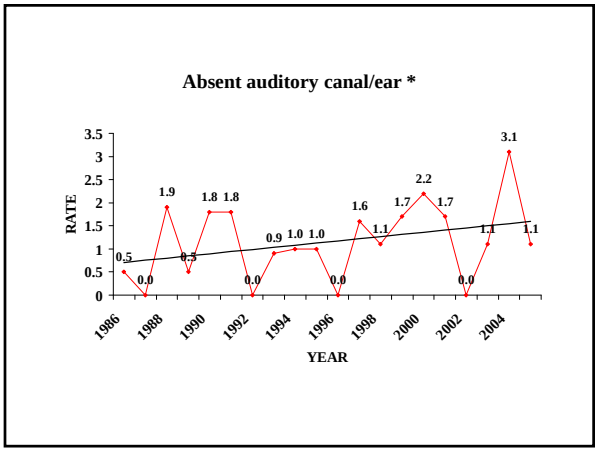

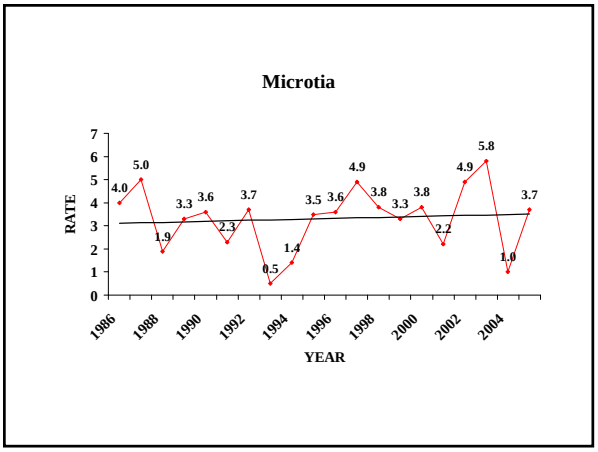

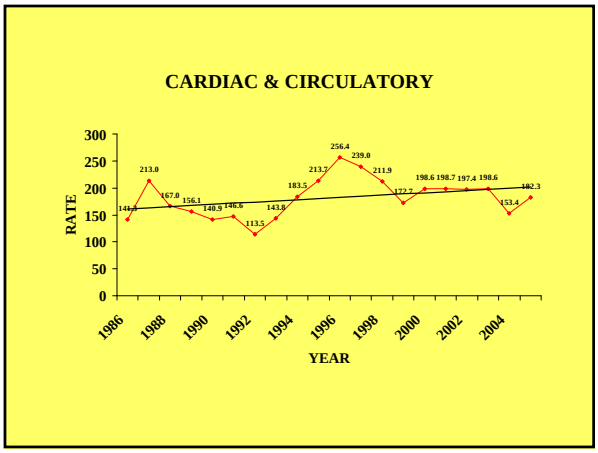

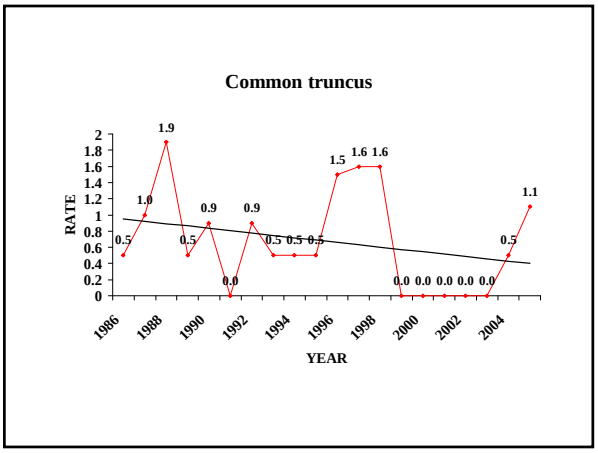

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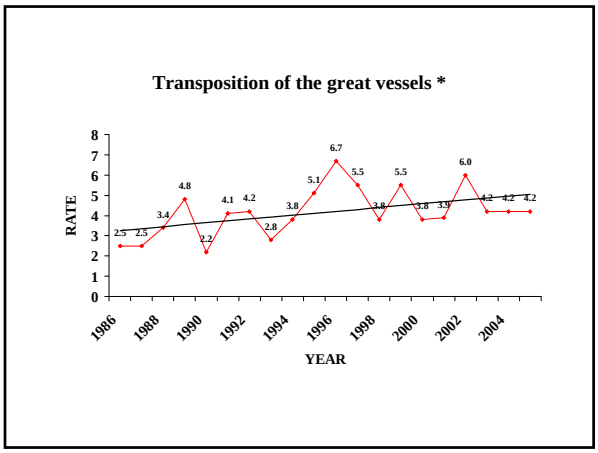

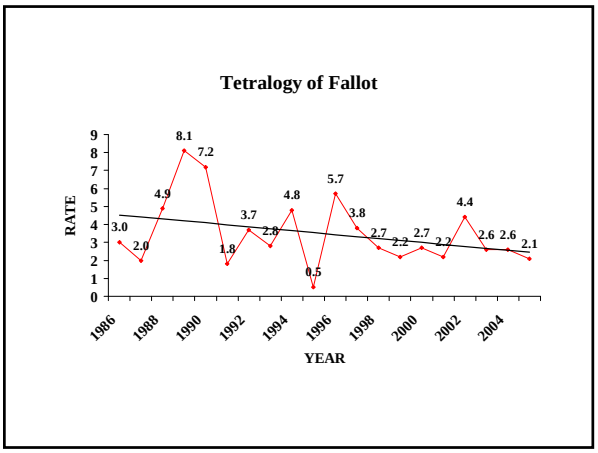

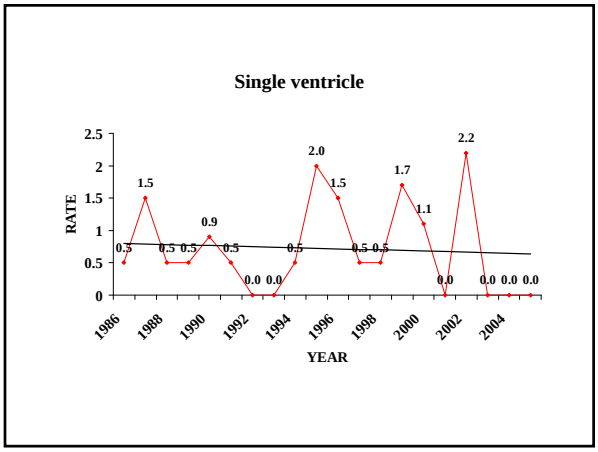

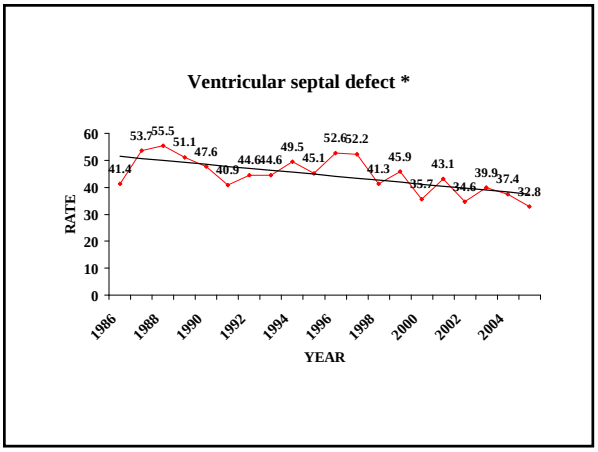

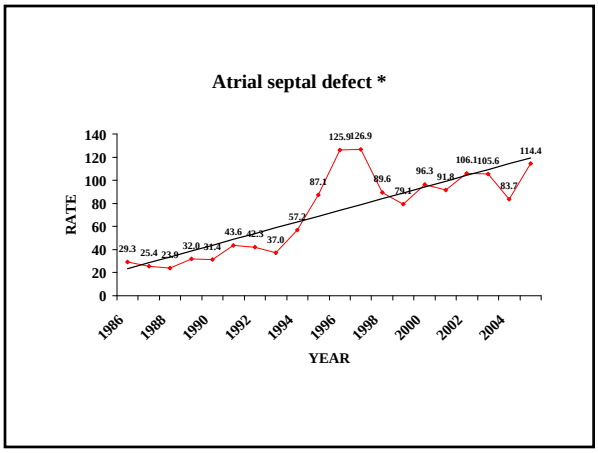

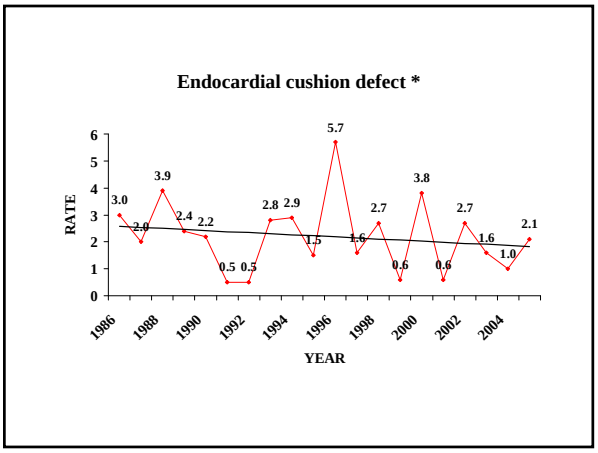

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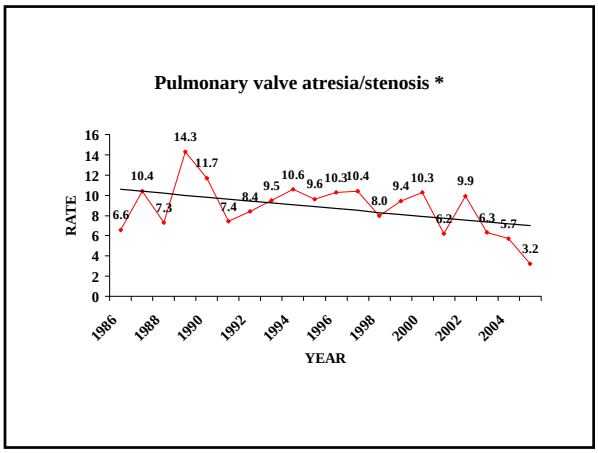

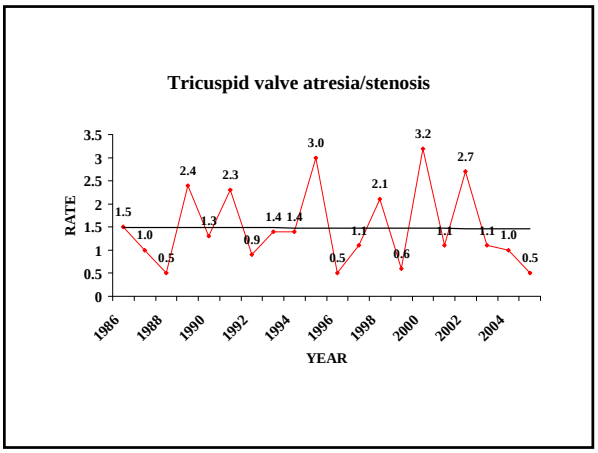

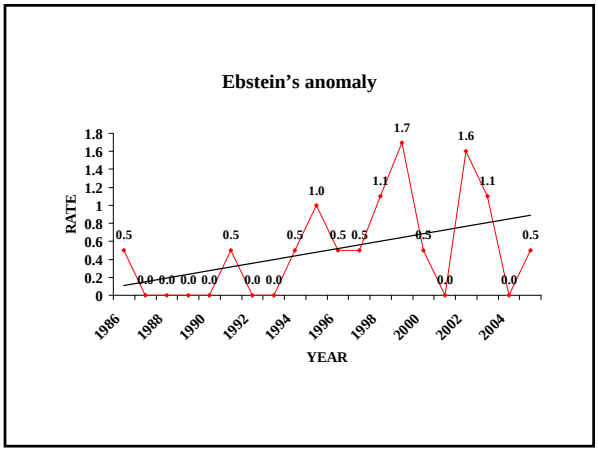

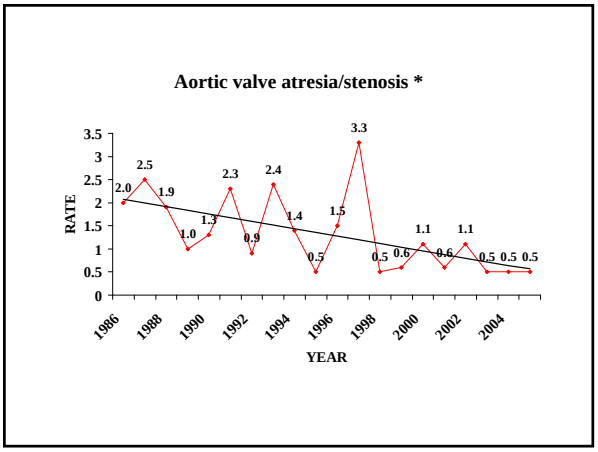

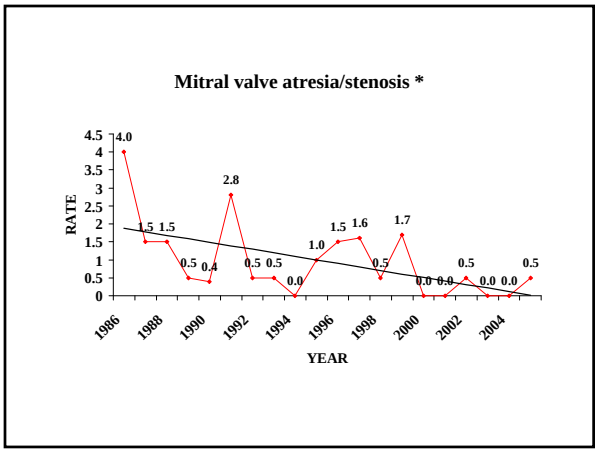

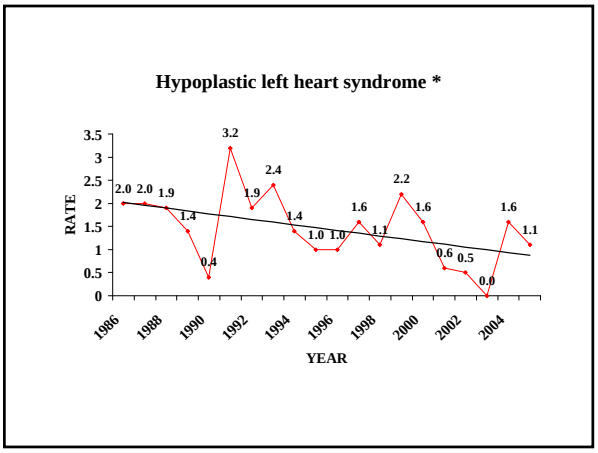

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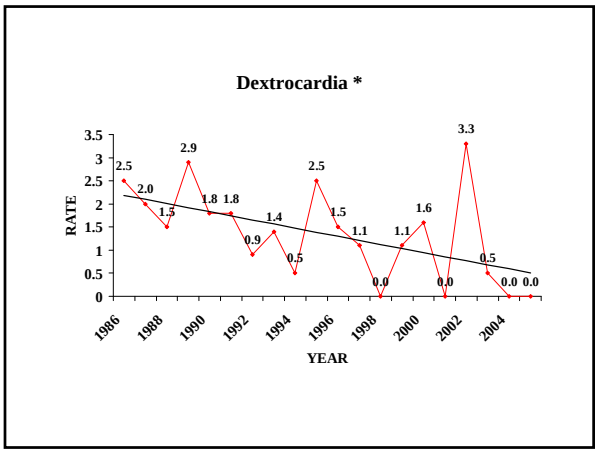

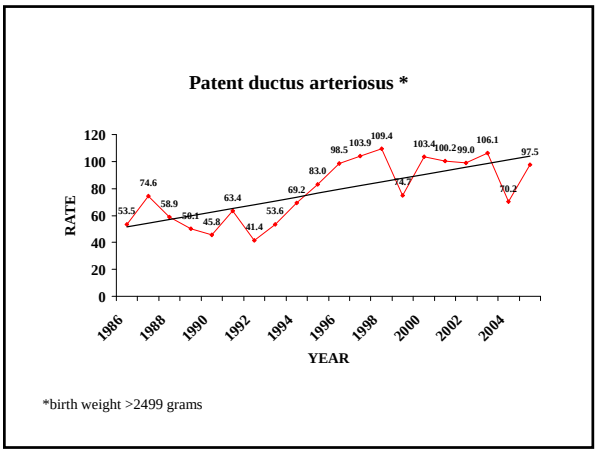

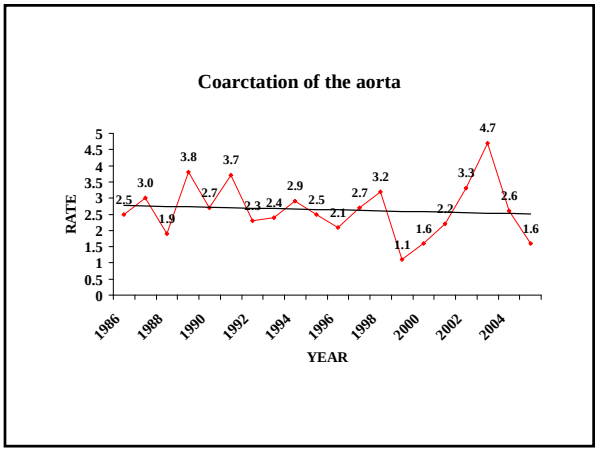

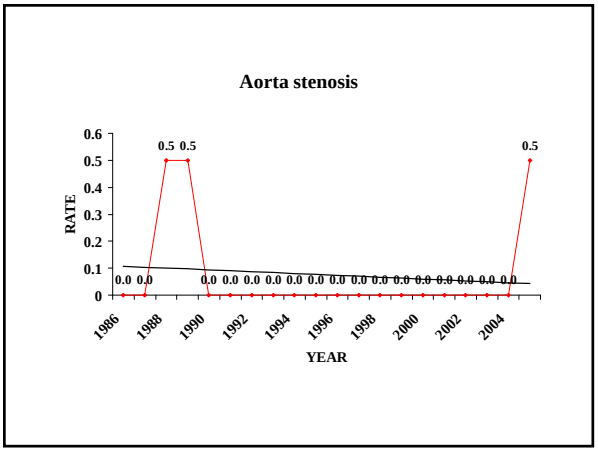

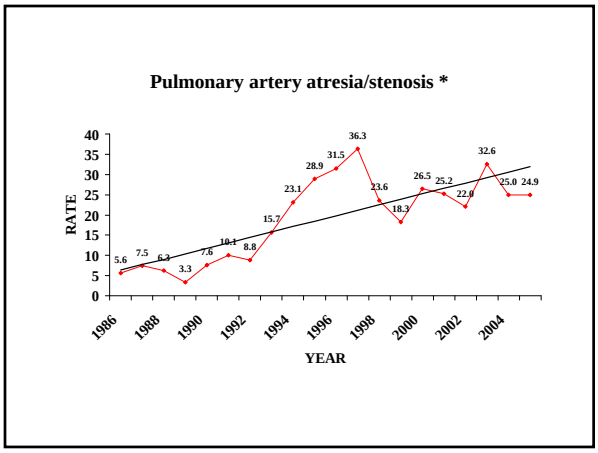

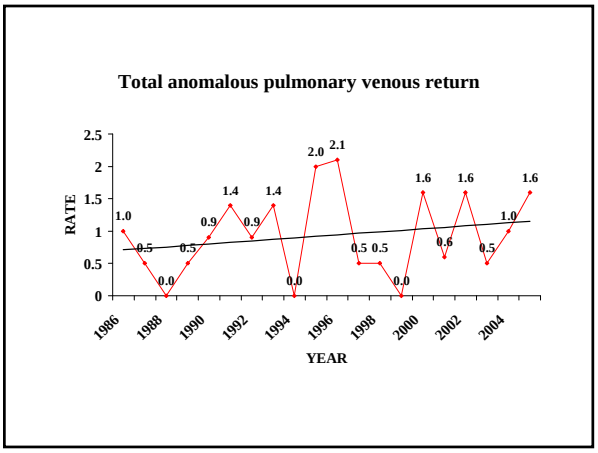

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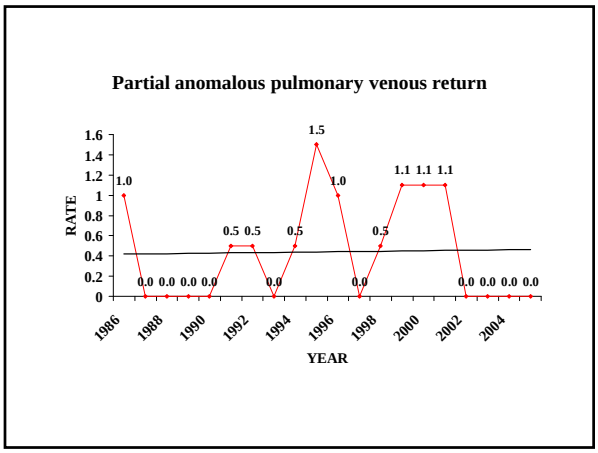

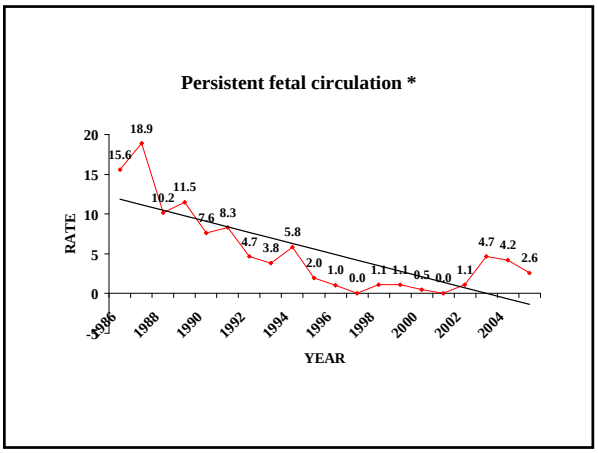

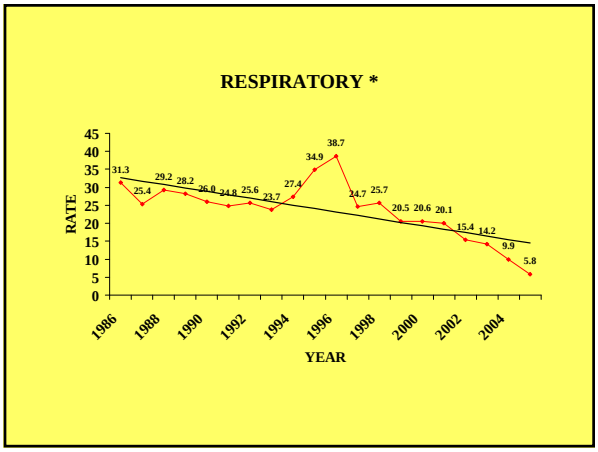

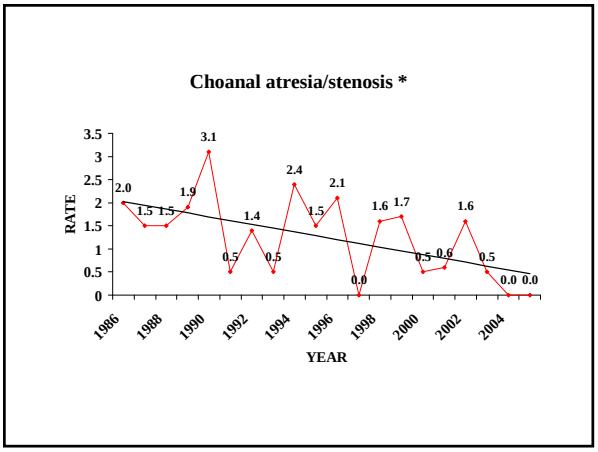

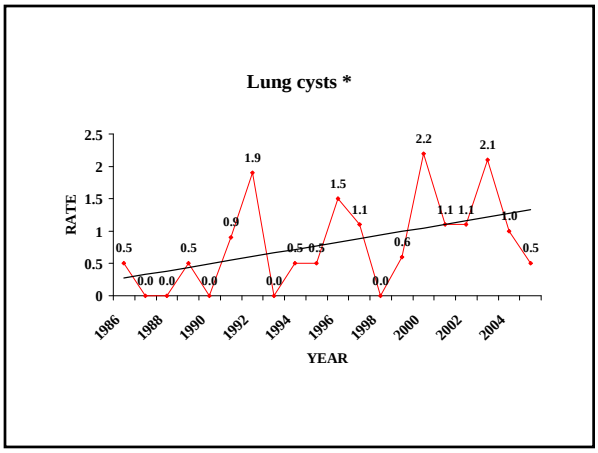

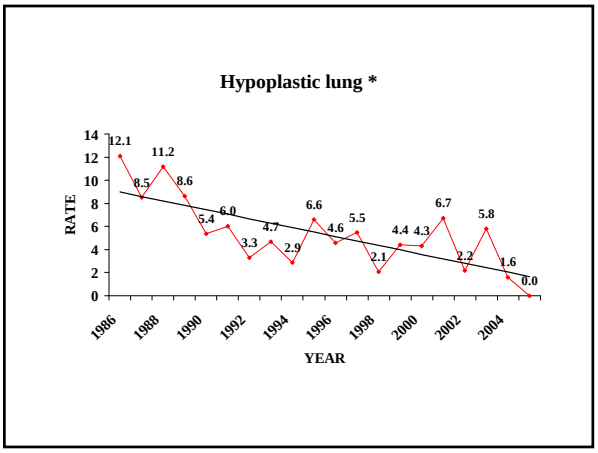

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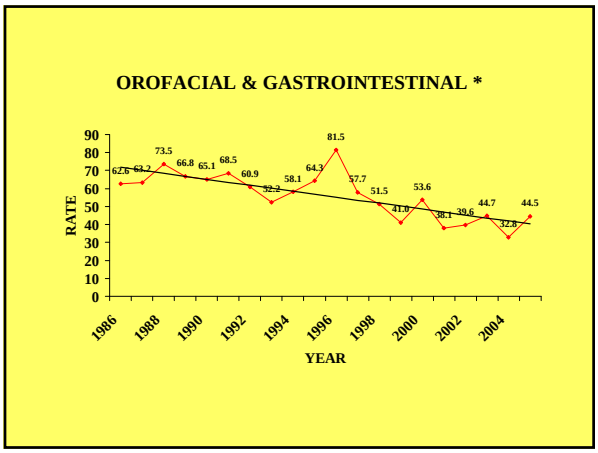

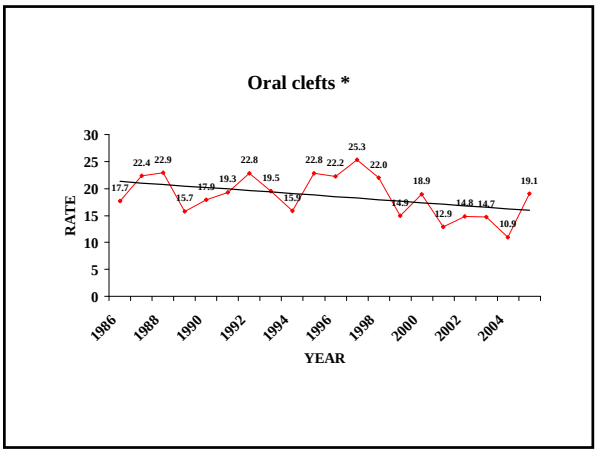

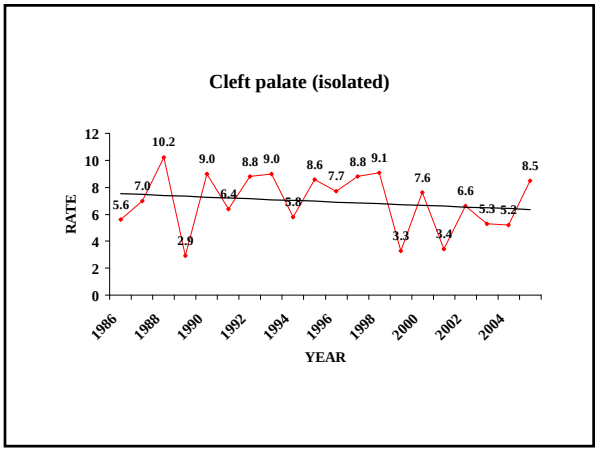

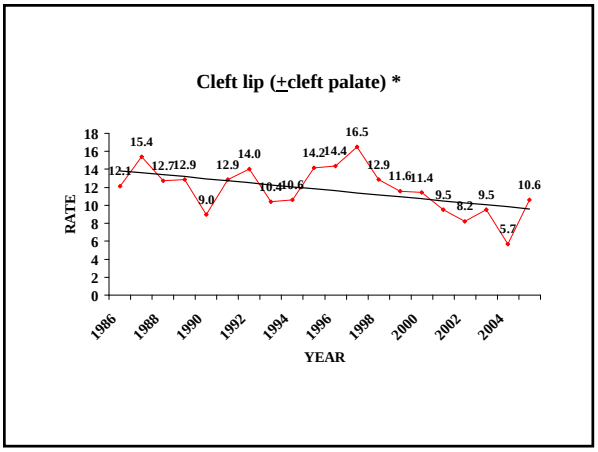

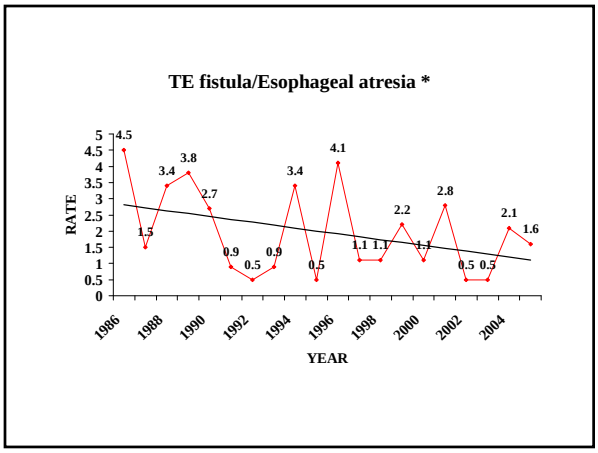

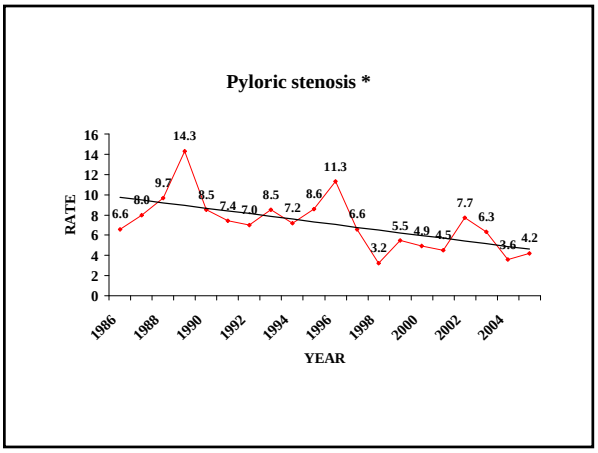

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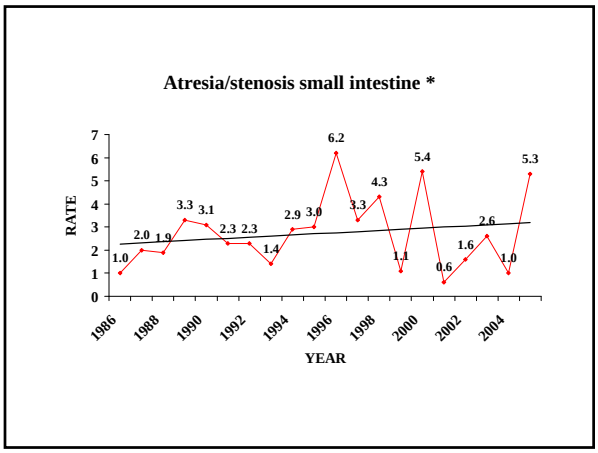

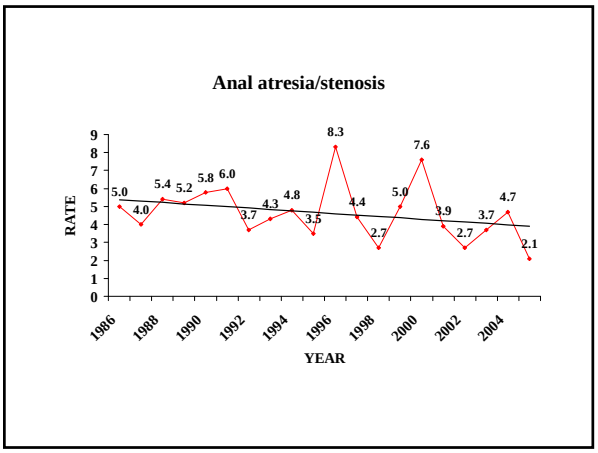

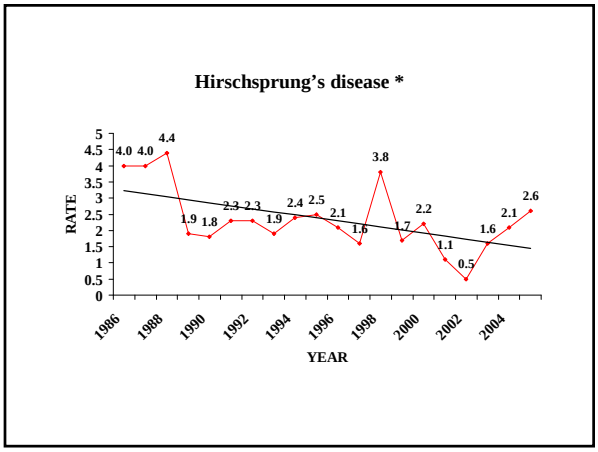

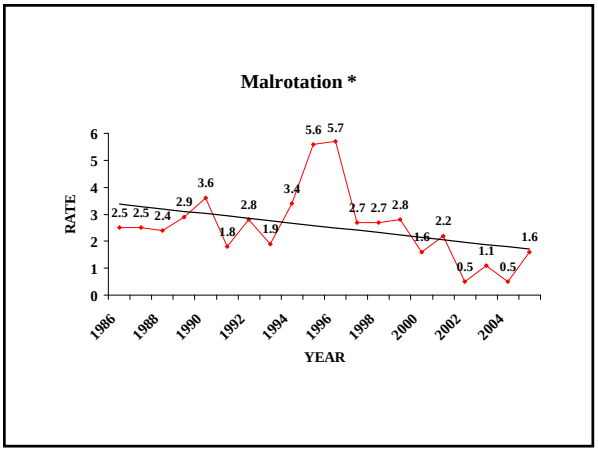

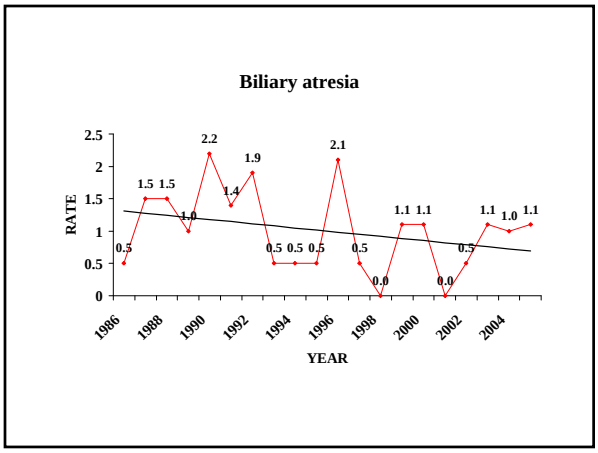

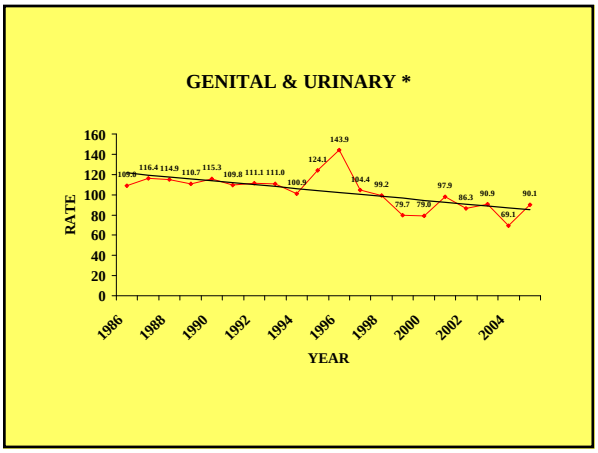

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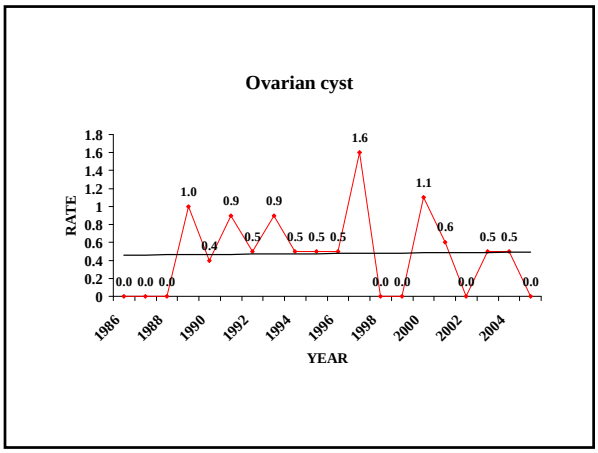

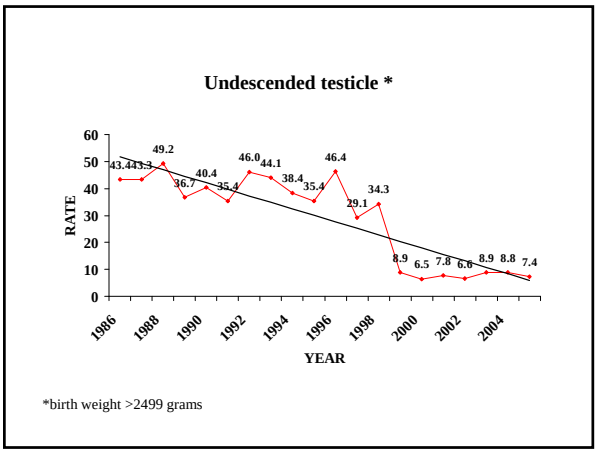

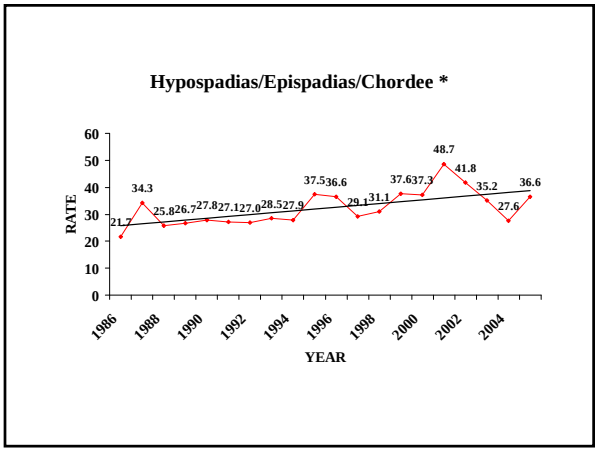

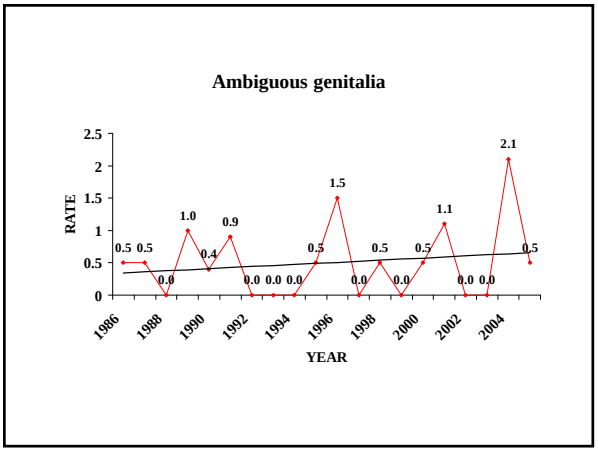

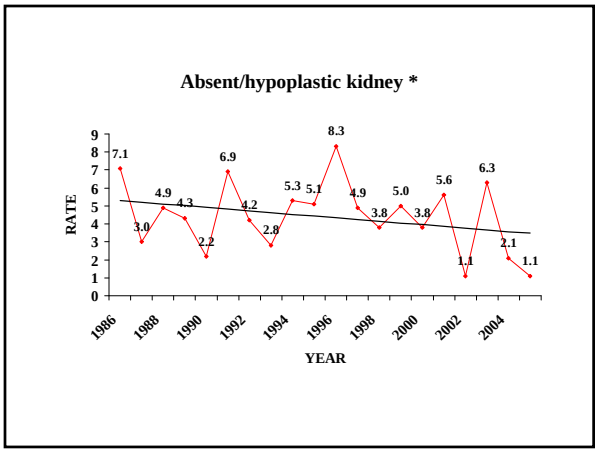

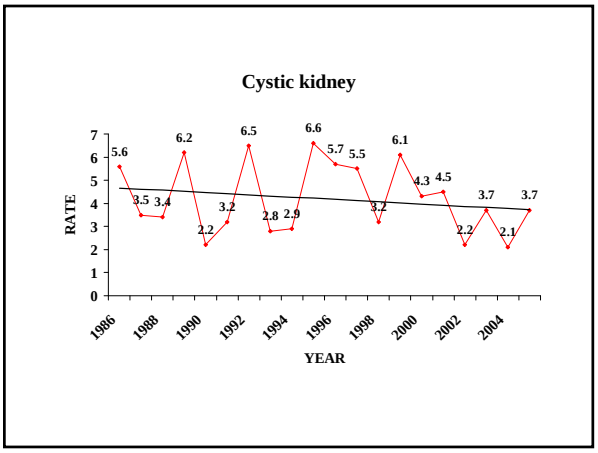

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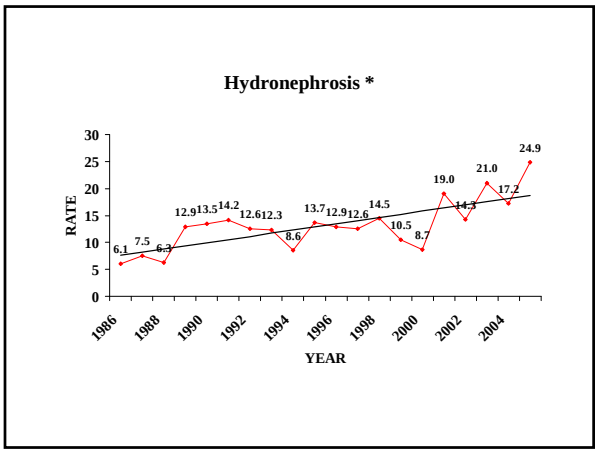

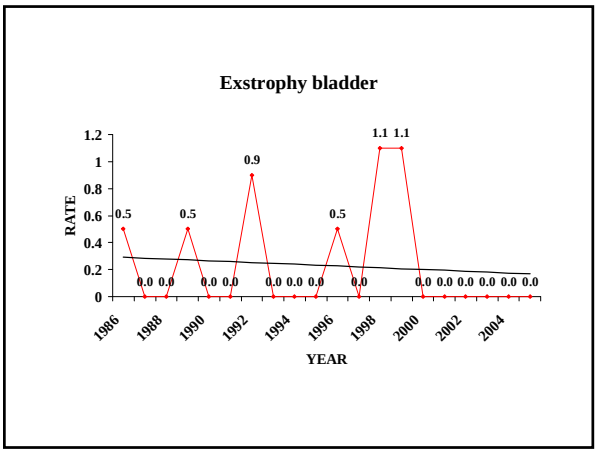

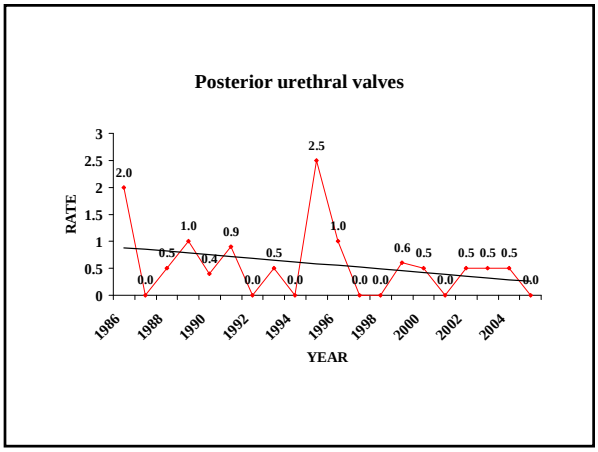

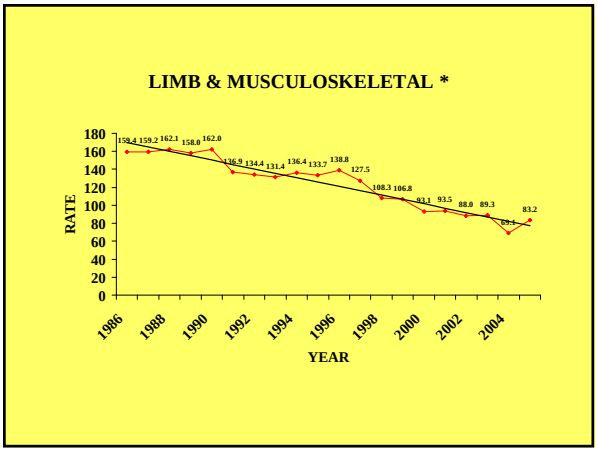

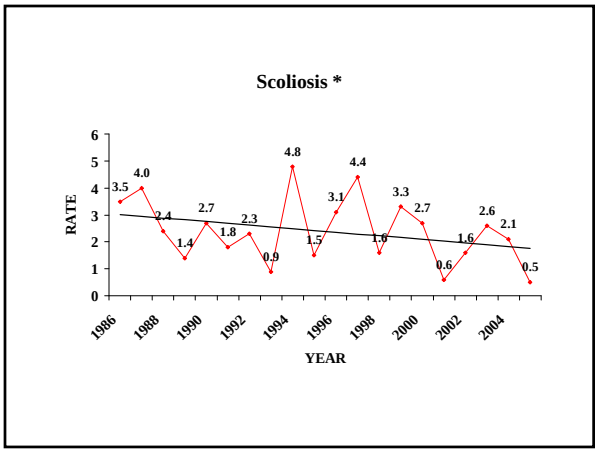

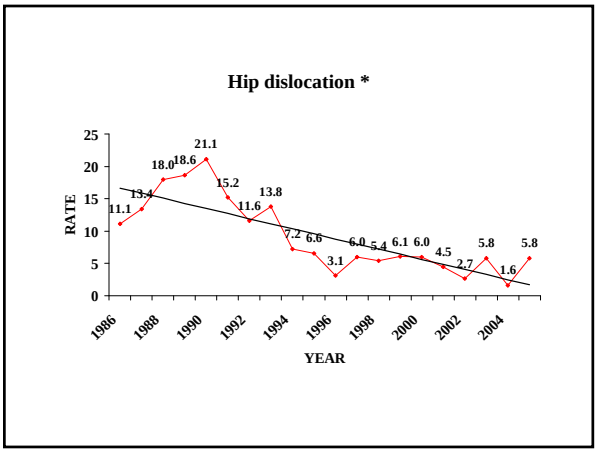

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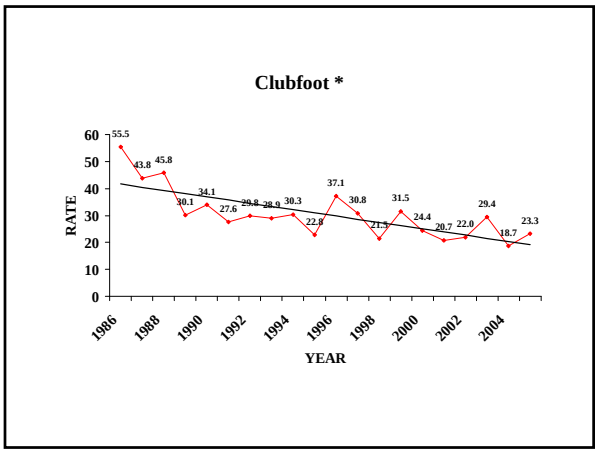

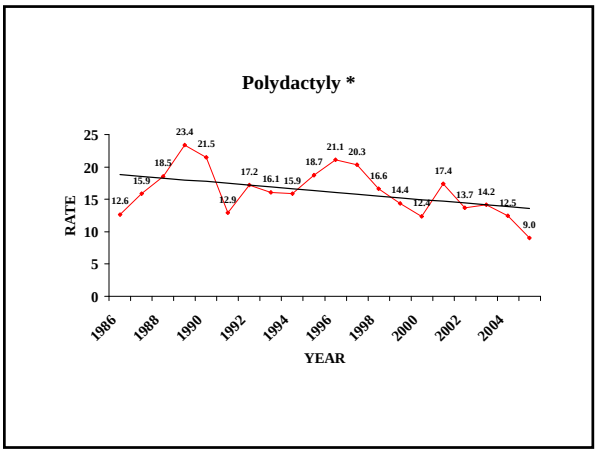

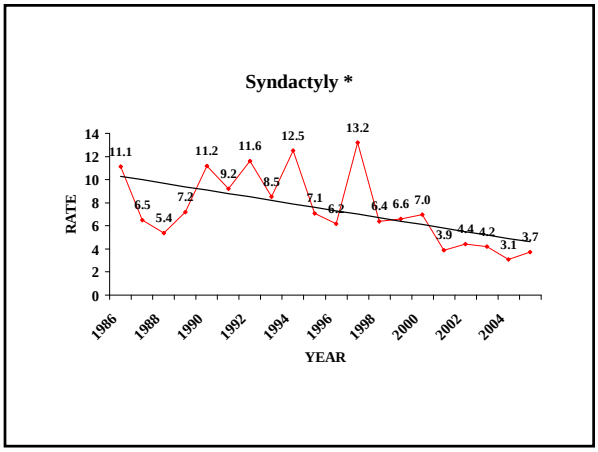

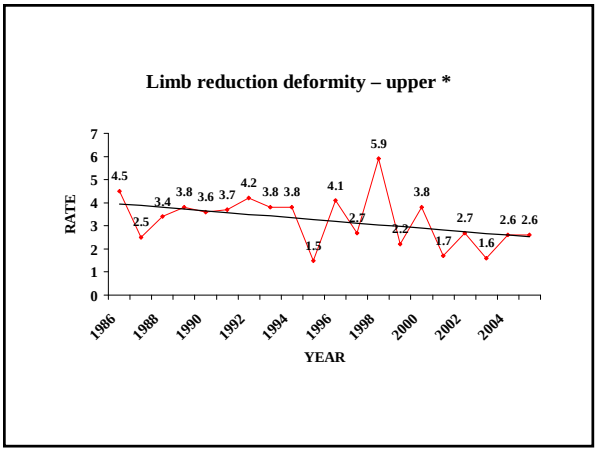

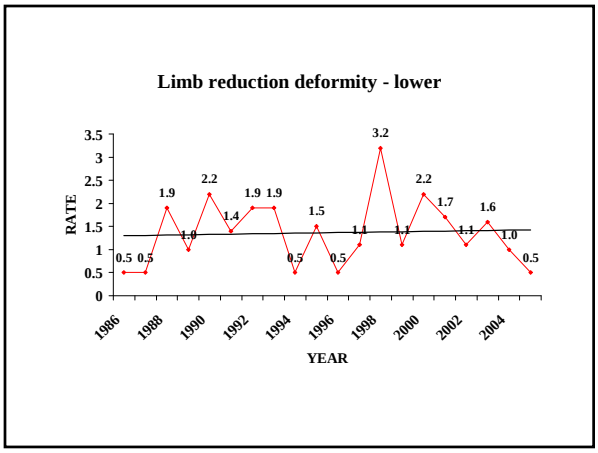

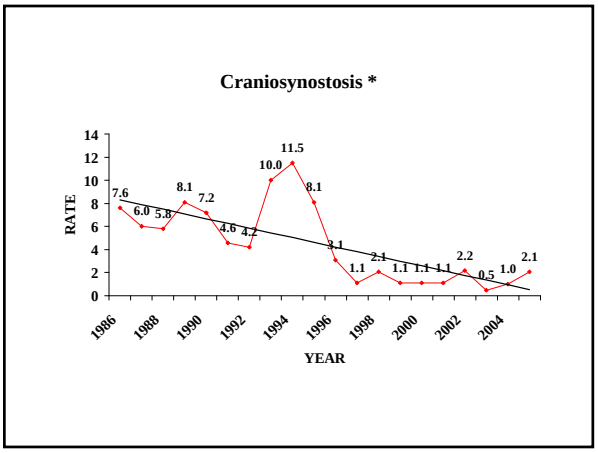

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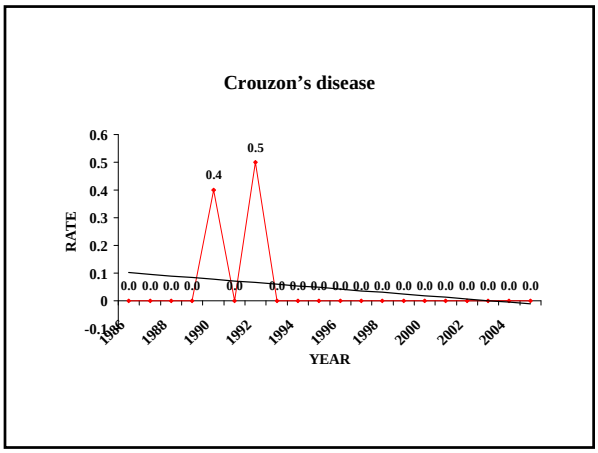

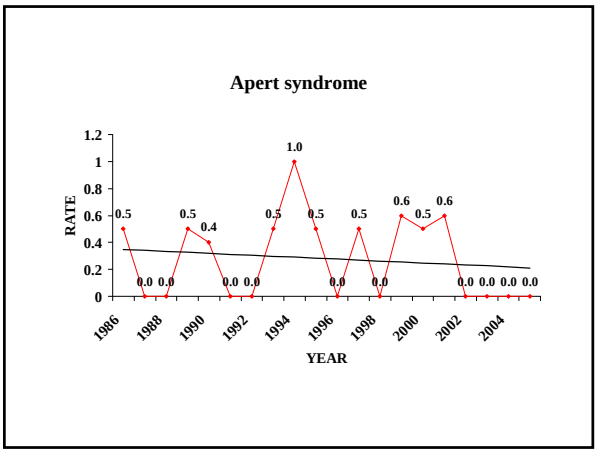

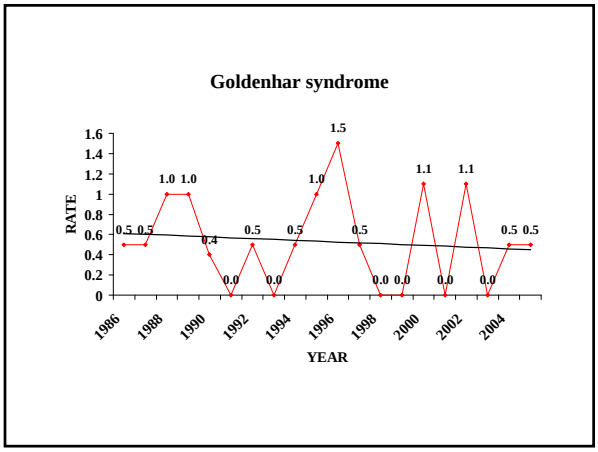

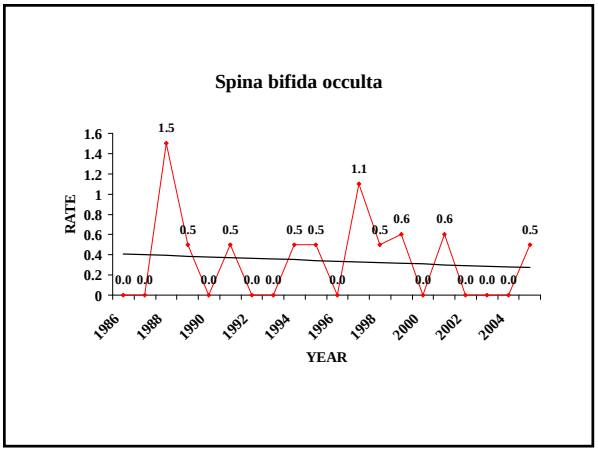

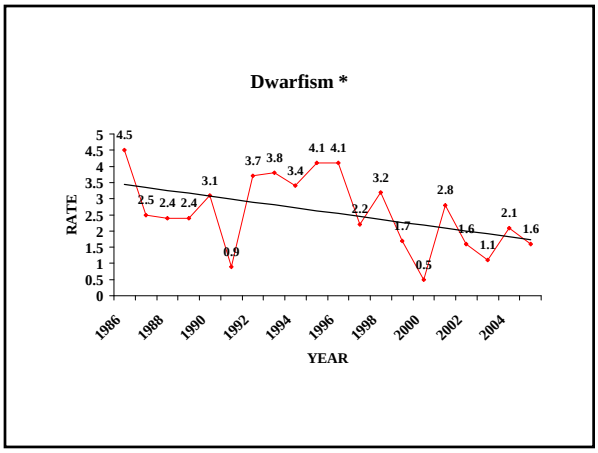

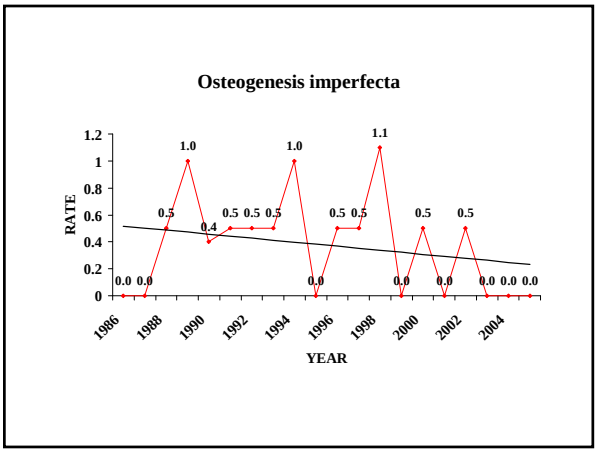

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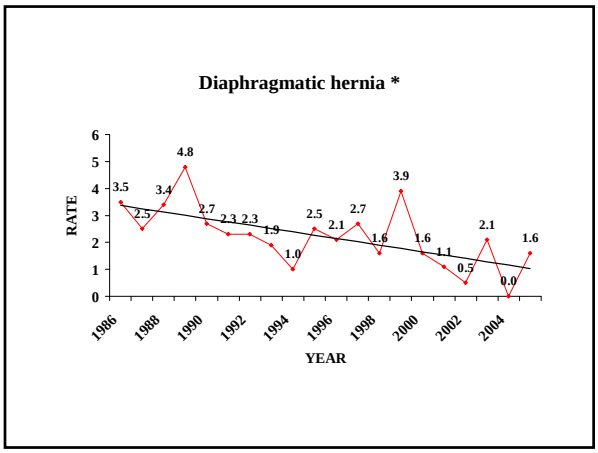

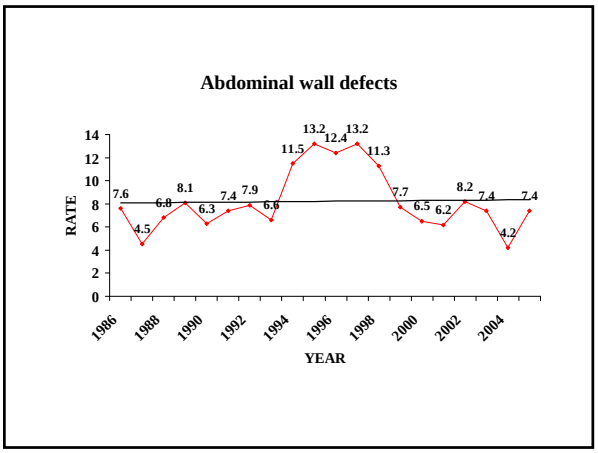

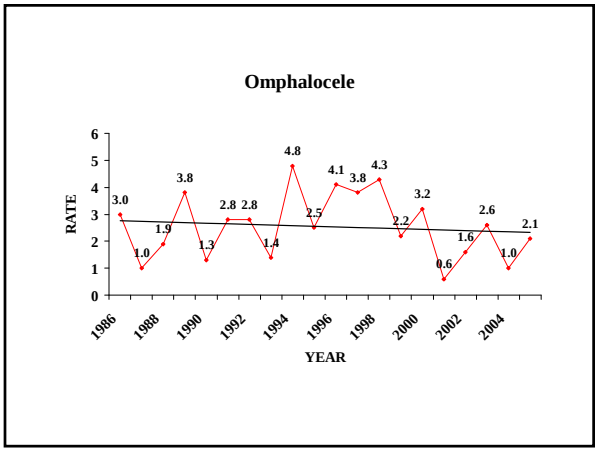

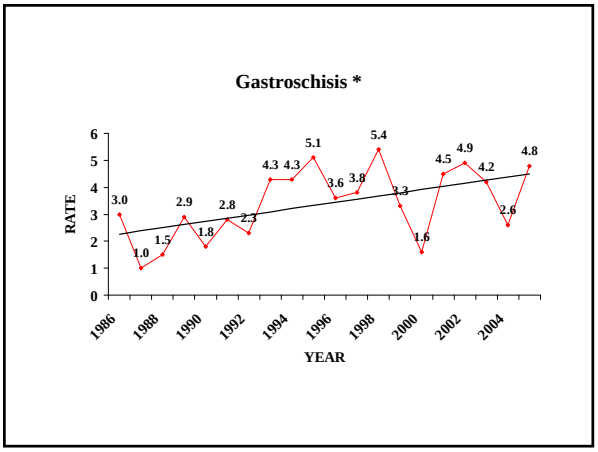

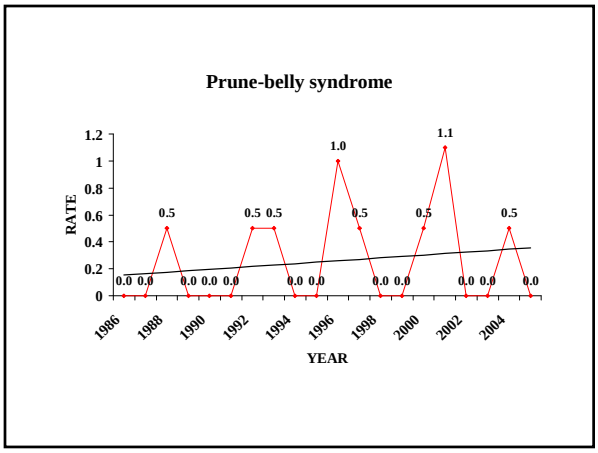

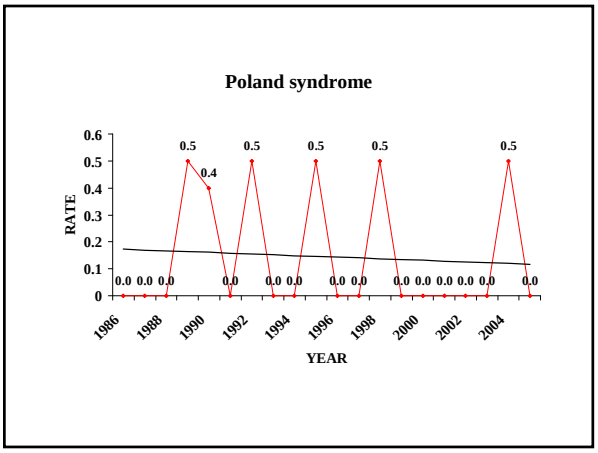

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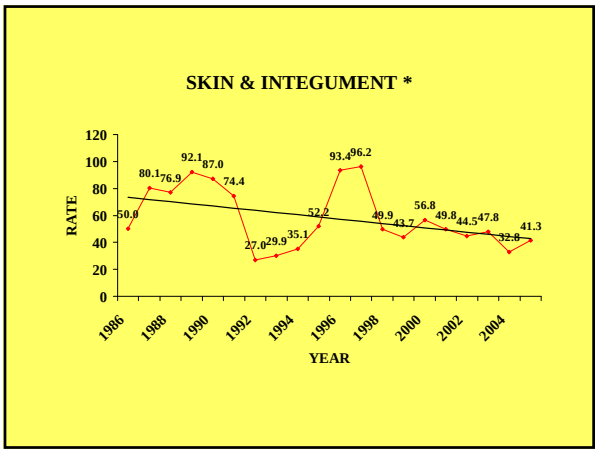

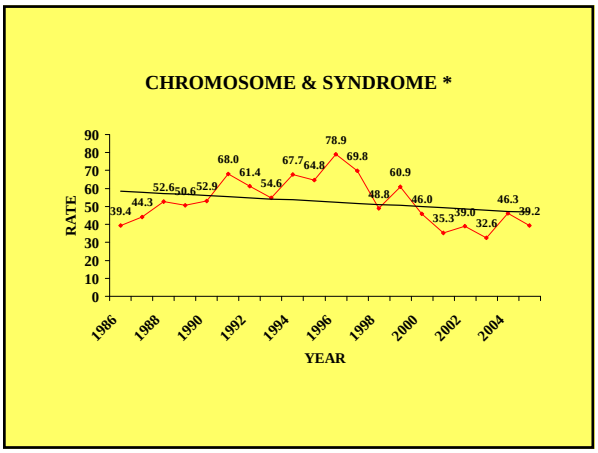

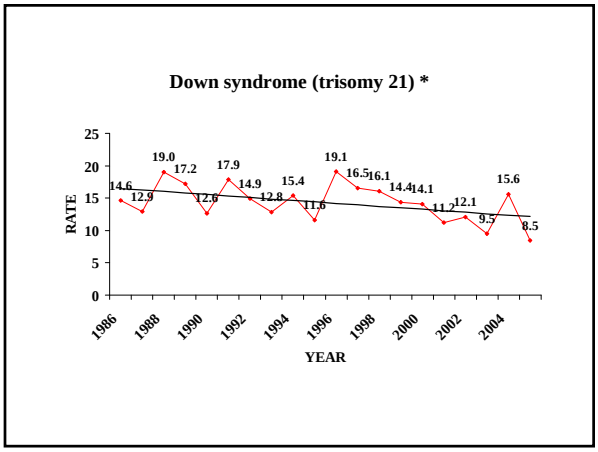

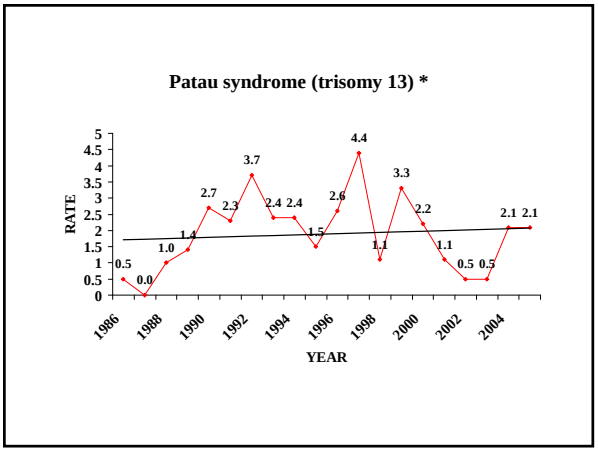

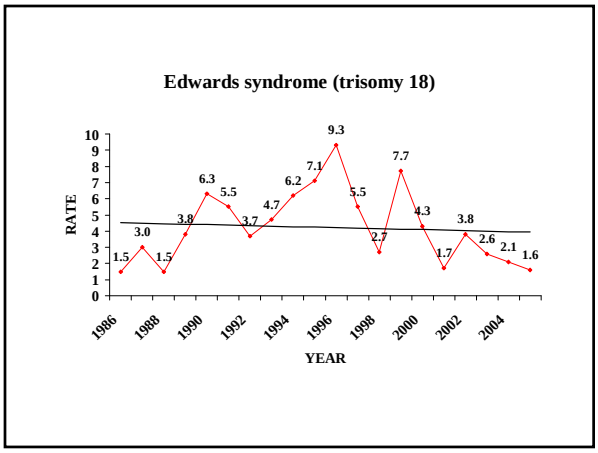

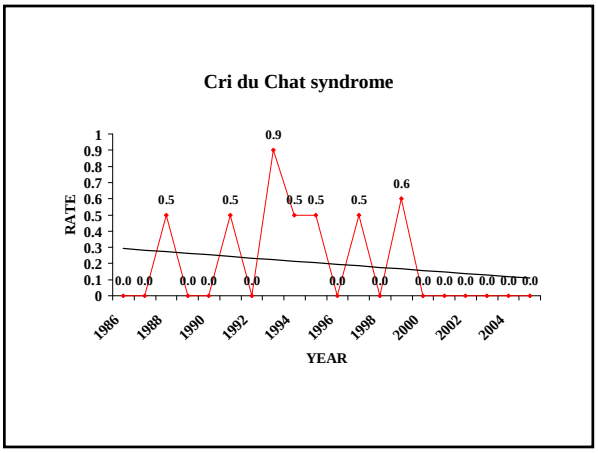

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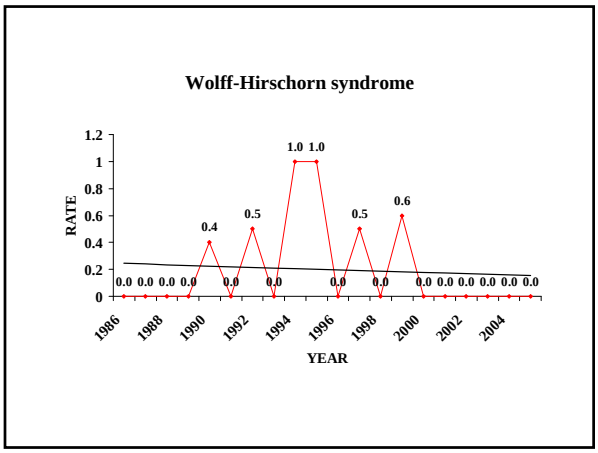

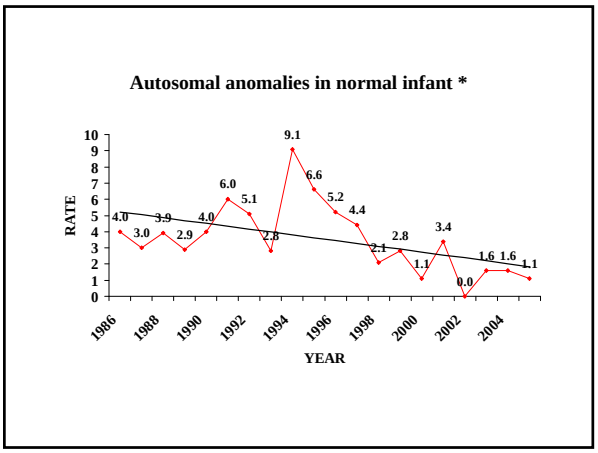

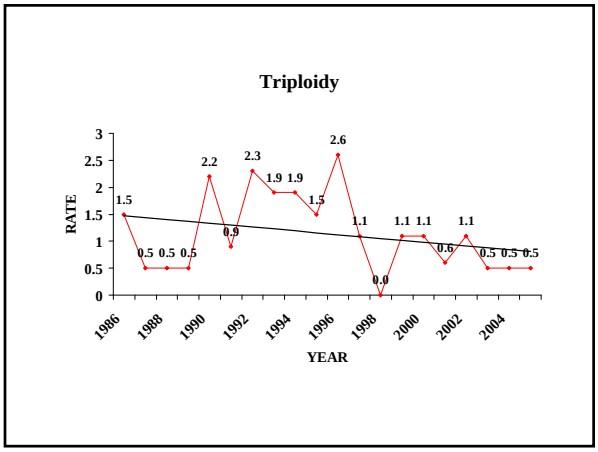

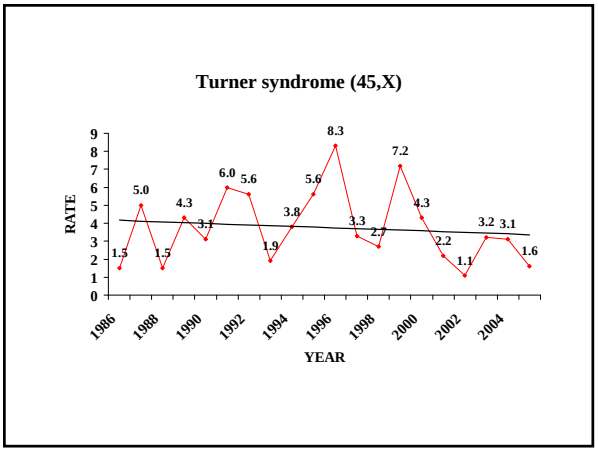

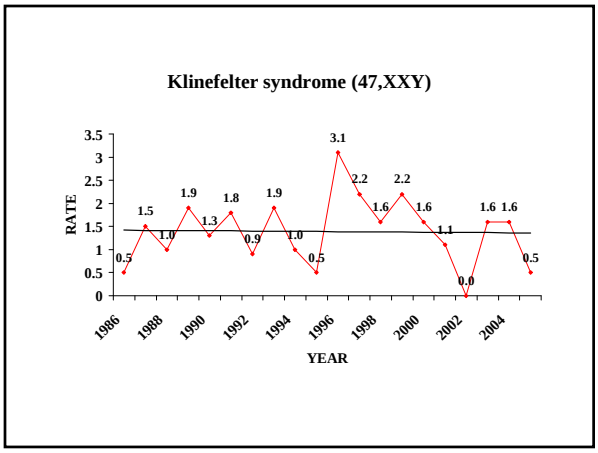

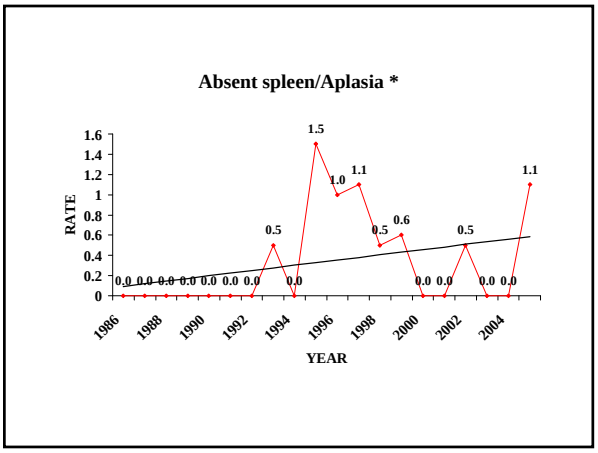

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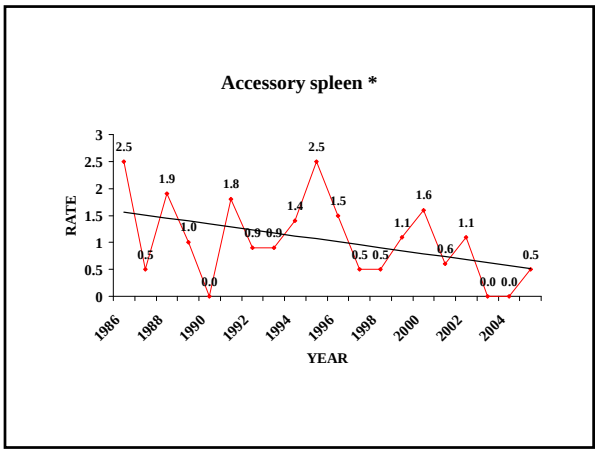

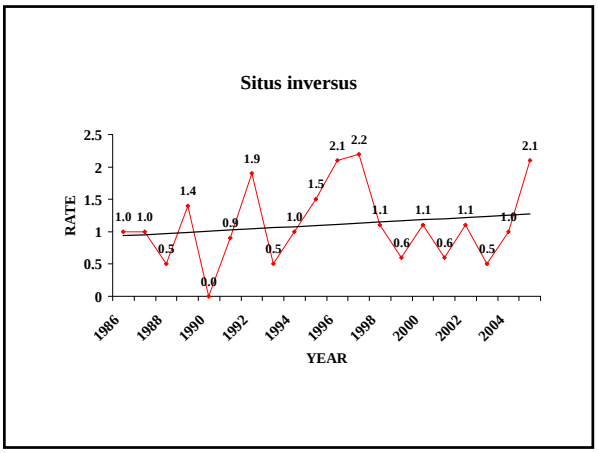

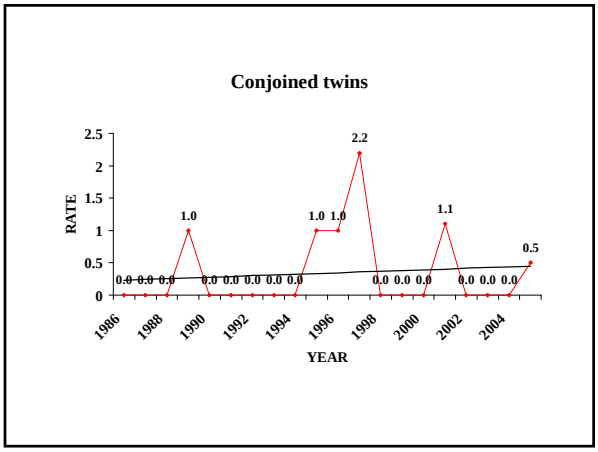

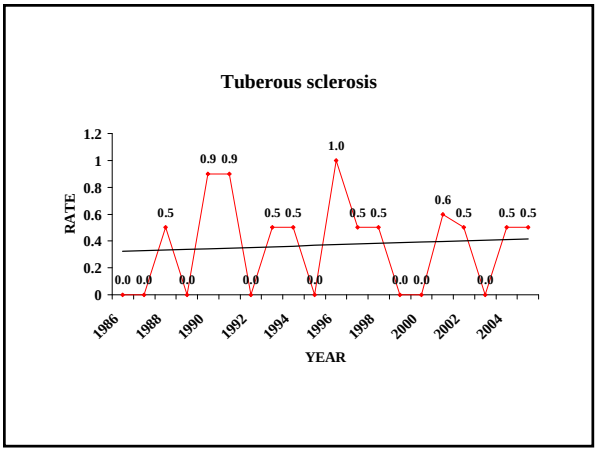

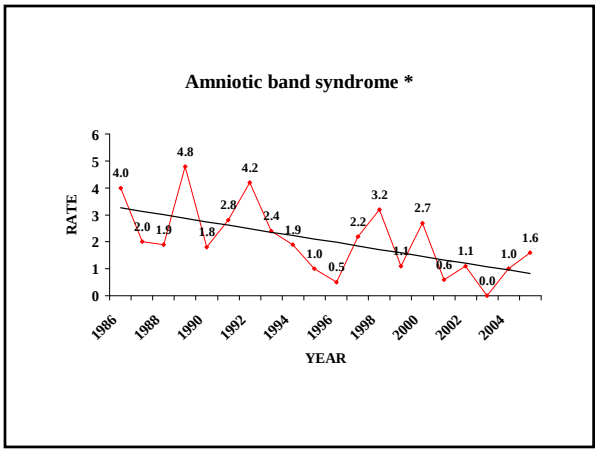

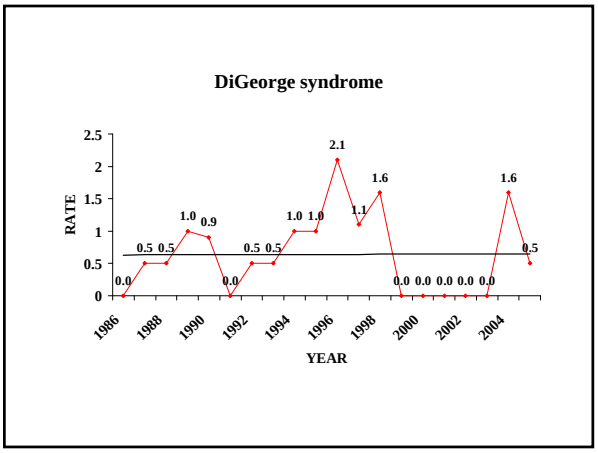

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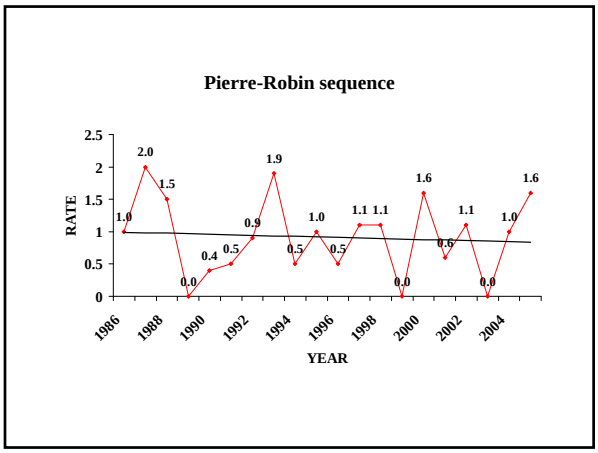

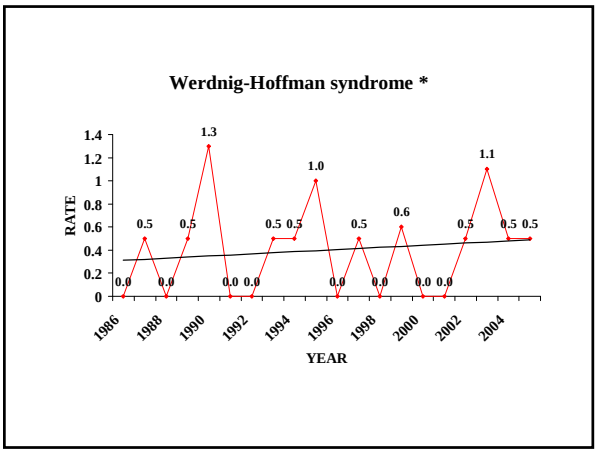

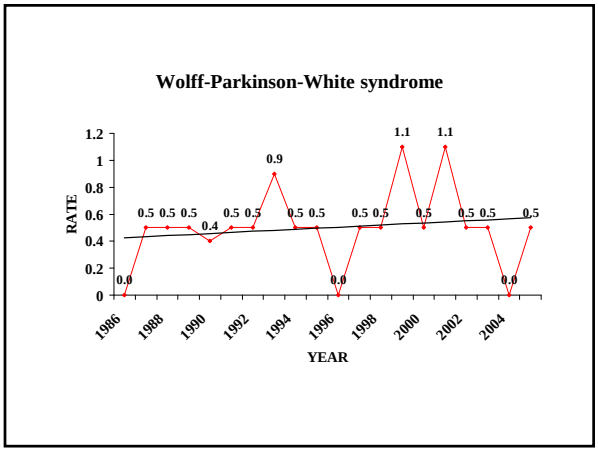

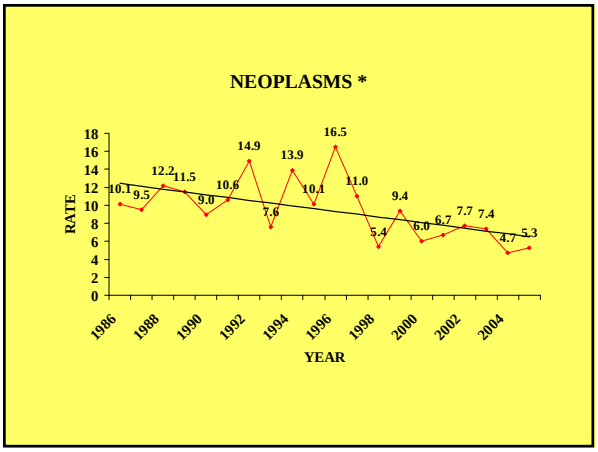

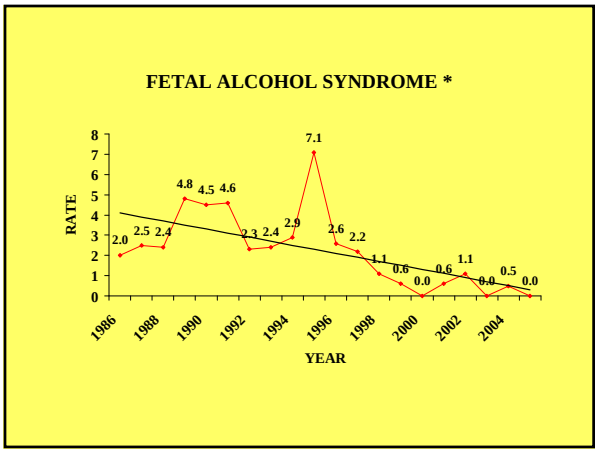

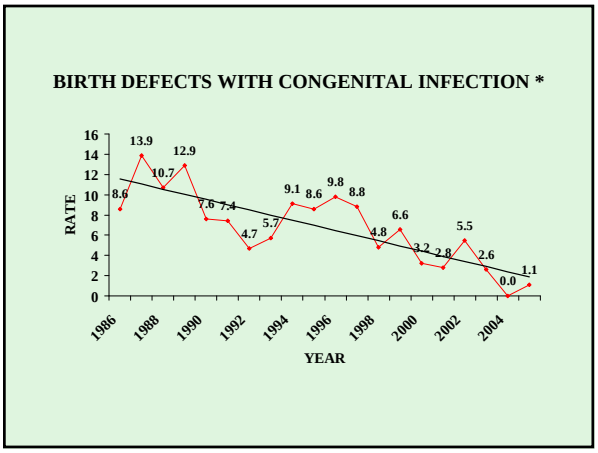

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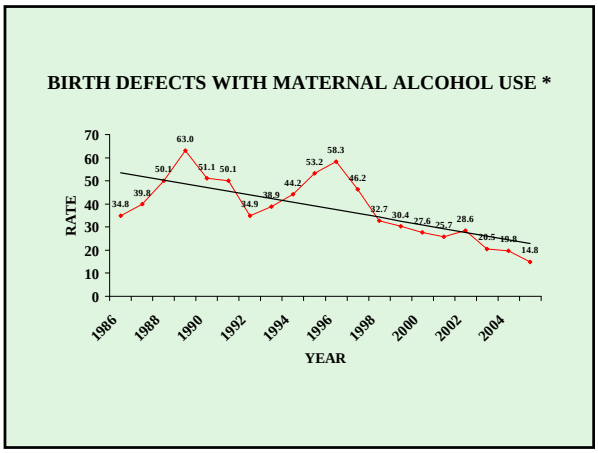

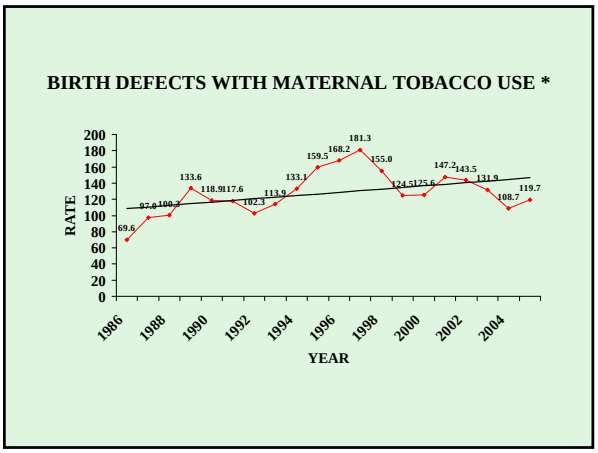

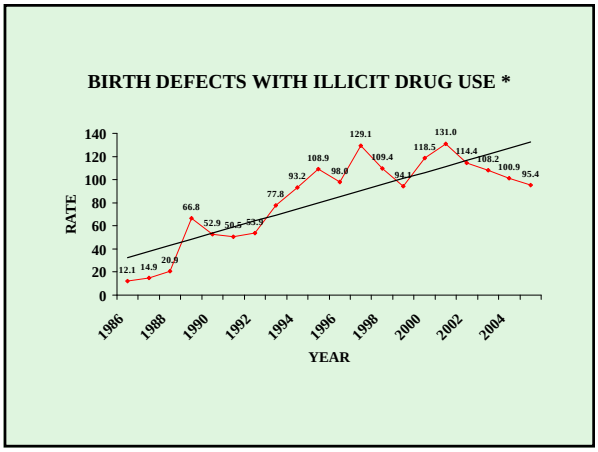

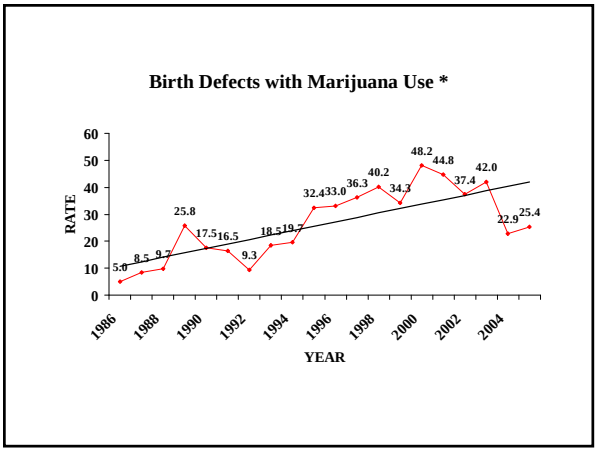

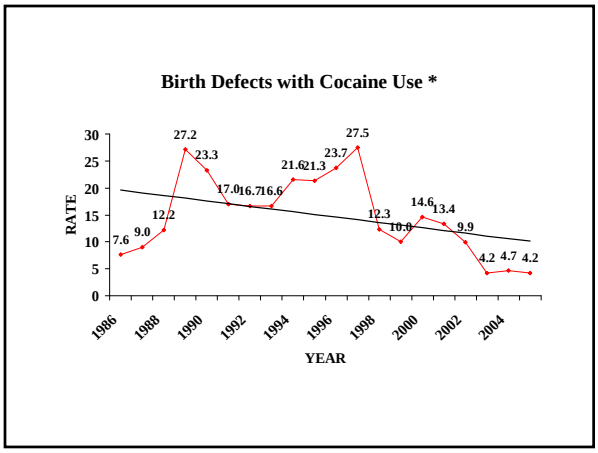

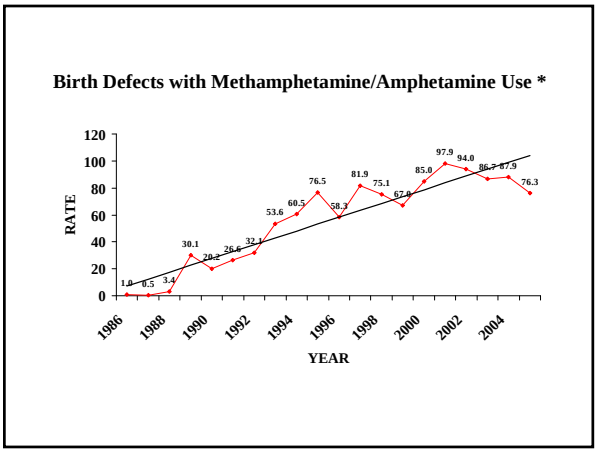

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TABLE 2. RATE OF BIRTH DEFECTS PER 10,000 TOTAL BIRTHS* BY YEAR, HAWAI'I, 1986-2005

Diagnosis	Total 1986-2005 N=394,569*		1986 N=19,819		1987 N=20,097		1988 N=20,541		1989 N=20,954		1990 N=22,288		1991 N=21,765	
	N**	Rate***	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
BRAIN & NERVOUS SYSTEM	1,543	39.1	74	37.3	74	36.8	73	35.5	86	41.0	85	38.1	83	38.1
Neural tube defects	358	9.1	16	8.1	18	9.0	13	6.3	21	10.0	18	8.1	11	5.1
Anencephaly	133	3.4	5	2.5	11	5.5	5	2.4	5	2.4	8	3.6	5	2.3
Spina bifida	162	4.1	9	4.5	5	2.5	6	2.9	10	4.8	5	2.2	4	1.8
Encephalocele	73	1.9	3	1.5	2	1.0	3	1.5	7	3.3	5	2.2	3	1.4
Microcephaly	370	9.4	27	13.6	24	11.9	25	12.2	28	13.4	17	7.6	27	12.4
Holoprosencephaly	48	1.2	1	0.5	2	1.0	1	0.5	2	1.0	3	1.3	2	0.9
Hydrocephaly (w/o spina bifida)	404	10.2	26	13.1	21	10.4	20	9.7	20	9.5	30	13.5	26	11.9
EYE, EAR, FACE, & NECK	1,895	48.0	105	53.0	108	53.7	109	53.1	116	55.4	110	49.4	117	53.8
EYE	711	18.0	34	17.2	37	18.4	49	23.9	49	23.4	48	21.5	48	22.1
Anophthalmia	14	0.4	0	0.0	1	0.5	0	0.0	1	0.5	1	0.4	0	0.0
Microphthalmia	109	2.8	2	1.0	7	3.5	8	3.9	10	4.8	9	4.0	8	3.7
Cataract	45	1.1	0	0.0	3	1.5	5	2.4	3	1.4	1	0.4	2	0.9
EAR	1,333	33.8	85	42.9	74	36.8	81	39.4	87	41.5	82	36.8	84	38.6
Absent auditory canal/ear	45	1.1	1	0.5	0	0.0	4	1.9	1	0.5	4	1.8	4	1.8
Microtia	130	3.3	8	4.0	10	5.0	4	1.9	7	3.3	8	3.6	5	2.3
CARDIAC & CIRCULATORY	7,086	179.6	280	141.3	428	213.0	343	167.0	327	156.1	314	140.9	319	146.6
Common truncus	27	0.7	1	0.5	2	1.0	4	1.9	1	0.5	2	0.9	0	0.0
Transposition of the great vessels	164	4.2	5	2.5	5	2.5	7	3.4	10	4.8	5	2.2	9	4.1
Tetralogy of Fallot	140	3.5	6	3.0	4	2.0	10	4.9	17	8.1	16	7.2	4	1.8
Single ventricle	28	0.7	1	0.5	3	1.5	1	0.5	1	0.5	2	0.9	1	0.5
Ventricular septal defect	1,761	44.6	82	41.4	108	53.7	114	55.5	107	51.1	106	47.6	89	40.9
Atrial septal defect	2,748	69.6	58	29.3	51	25.4	49	23.9	67	32.0	70	31.4	95	43.6
Endocardial cushion defect	87	2.2	6	3.0	4	2.0	8	3.9	5	2.4	5	2.2	1	0.5
Pulmonary valve atresia/stenosis	348	8.8	13	6.6	21	10.4	15	7.3	30	14.3	26	11.7	16	7.4
Tricuspid valve atresia/stenosis	59	1.5	3	1.5	2	1.0	1	0.5	5	2.4	3	1.3	5	2.3
Ebstein's anomaly	19	0.5	1	0.5	0	0.0	0	0.0	0	0.0	0	0.0	1	0.5
Aortic valve atresia/stenosis	53	1.3	4	2.0	5	2.5	4	1.9	2	1.0	3	1.3	5	2.3
Mitral valve atresia/stenosis	38	1.0	8	4.0	3	1.5	3	1.5	1	0.5	1	0.4	6	2.8
Hypoplastic left heart syndrome	58	1.5	4	2.0	4	2.0	4	1.9	3	1.4	1	0.4	7	3.2
Dextrocardia	53	1.3	5	2.5	4	2.0	3	1.5	6	2.9	4	1.8	4	1.8
Patent ductus arteriosus +	3,023	76.6	106	53.5	150	74.6	121	58.9	105	50.1	102	45.8	138	63.4
Coarctation of the aorta	105	2.7	5	2.5	6	3.0	4	1.9	8	3.8	6	2.7	8	3.7

*Total births=live births+fetal deaths **N=live births, fetal deaths, and terminations with birth defects ***Rate=(number of birth defects)/(total births)x10,000 +Birth weight >2,499 grams

TABLE 2. RATE OF BIRTH DEFECTS PER 10,000 TOTAL BIRTHS* BY YEAR, HAWAI'I, 1986-2005

Diagnosis	1992		1993		1994		1995		1996		1997		1998	
	N=21,504		N=21,073		N=20,814		N=19,750		N=19,386		N=18,198		N=19,644	
	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
BRAIN & NERVOUS SYSTEM	87	40.5	100	47.5	96	46.1	89	45.1	118	60.9	81	44.5	62	33.3
Neural tube defects	30	14.0	26	12.3	27	13.0	20	10.1	37	19.1	17	9.3	12	6.4
Anencephaly	10	4.7	8	3.8	9	4.3	6	3.0	10	5.2	8	4.4	6	3.2
Spina bifida	17	7.9	15	7.1	12	5.8	11	5.6	14	7.2	9	4.9	5	2.7
Encephalocele	3	1.4	3	1.4	7	3.4	3	1.5	14	7.2	0	0.0	4	2.1
Microcephaly	17	7.9	26	12.3	18	8.6	25	12.7	21	10.8	17	9.3	11	5.9
Holoprosencephaly	1	0.5	1	0.5	7	3.4	4	2.0	2	1.0	0	0.0	3	1.6
Hydrocephaly (w/o spina bifida)	21	9.8	17	8.1	28	13.5	26	13.2	29	15.0	22	12.1	20	10.7
EYE, EAR, FACE, & NECK	96	44.6	89	42.2	106	50.9	125	63.3	117	60.4	102	56.1	96	51.5
EYE	43	20.0	34	16.1	44	21.1	49	24.8	51	26.3	38	20.9	39	20.9
Anophthalmia	0	0.0	2	0.9	1	0.5	0	0.0	1	0.5	2	1.1	0	0.0
Microphthalmia	4	1.9	6	2.8	6	2.9	12	6.1	7	3.6	6	3.3	3	1.6
Cataract	4	1.9	1	0.5	2	1.0	6	3.0	3	1.5	5	2.7	1	0.5
EAR	66	30.7	63	29.9	73	35.1	91	46.1	84	43.3	69	37.9	60	32.2
Absent auditory canal/ear	0	0.0	2	0.9	2	1.0	2	1.0	0	0.0	3	1.6	2	1.1
Microtia	8	3.7	1	0.5	3	1.4	7	3.5	7	3.6	9	4.9	7	3.8
CARDIAC & CIRCULATORY	244	113.5	303	143.8	382	183.5	422	213.7	497	256.4	435	239.0	395	211.9
Common truncus	2	0.9	1	0.5	1	0.5	1	0.5	3	1.5	3	1.6	3	1.6
Transposition of the great vessels	9	4.2	6	2.8	8	3.8	10	5.1	13	6.7	10	5.5	7	3.8
Tetralogy of Fallot	8	3.7	6	2.8	10	4.8	1	0.5	11	5.7	7	3.8	5	2.7
Single ventricle	0	0.0	0	0.0	1	0.5	4	2.0	3	1.5	1	0.5	1	0.5
Ventricular septal defect	96	44.6	94	44.6	103	49.5	89	45.1	102	52.6	95	52.2	77	41.3
Atrial septal defect	91	42.3	78	37.0	119	57.2	172	87.1	244	125.9	231	126.9	167	89.6
Endocardial cushion defect	1	0.5	6	2.8	6	2.9	3	1.5	11	5.7	3	1.6	5	2.7
Pulmonary valve atresia/stenosis	18	8.4	20	9.5	22	10.6	19	9.6	20	10.3	19	10.4	15	8.0
Tricuspid valve atresia/stenosis	2	0.9	3	1.4	3	1.4	6	3.0	1	0.5	2	1.1	4	2.1
Ebstein's anomaly	0	0.0	0	0.0	1	0.5	2	1.0	1	0.5	1	0.5	2	1.1
Aortic valve atresia/stenosis	2	0.9	5	2.4	3	1.4	1	0.5	3	1.5	6	3.3	1	0.5
Mitral valve atresia/stenosis	1	0.5	1	0.5	0	0.0	2	1.0	3	1.5	3	1.6	1	0.5
Hypoplastic left heart syndrome	4	1.9	5	2.4	3	1.4	2	1.0	2	1.0	3	1.6	2	1.1
Dextrocardia	2	0.9	3	1.4	1	0.5	5	2.5	3	1.5	2	1.1	0	0.0
Patent ductus arteriosus +	89	41.4	113	53.6	144	69.2	164	83.0	191	98.5	189	103.9	204	109.4
Coarctation of the aorta	5	2.3	5	2.4	6	2.9	5	2.5	4	2.1	5	2.7	6	3.2

*Total births=live births+fetal deaths **N=live births, fetal deaths, and terminations with birth defects ***Rate=(number of birth defects)/(total births)x10,000 +Birth weight >2,499 grams

TABLE 2. RATE OF BIRTH DEFECTS PER 10,000 TOTAL BIRTHS* BY YEAR, HAWAI'I, 1986-2005

Diagnosis	1999 N=18,067		2000 N=18,478		2001 N=17867		2002 N=18,182		2003 N=19,033		2004 N=19,234		2005 N=18,875	
	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
BRAIN & NERVOUS SYSTEM	51	28.2	75	40.6	65	36.4	72	39.6	53	27.8	66	34.3	53	28.1
Neural tube defects	15	8.3	15	8.1	16	9.0	13	7.1	8	4.2	10	5.2	15	7.9
Anencephaly	4	2.2	6	3.2	8	4.5	4	2.2	2	1.1	6	3.1	7	3.7
Spina bifida	9	5.0	6	3.2	5	2.8	7	3.8	5	2.6	2	1.0	6	3.2
Encephalocele	2	1.1	3	1.6	3	1.7	2	1.1	1	0.5	2	1.0	3	1.6
Microcephaly	6	3.3	19	10.3	17	9.5	9	4.9	12	6.3	15	7.8	9	4.8
Holoprosencephaly	3	1.7	5	2.7	2	1.1	2	1.1	1	0.5	6	3.1	0	0.0
Hydrocephaly (w/o spina bifida)	14	7.7	20	10.8	13	7.3	19	10.4	13	6.8	11	5.7	8	4.2
EYE, EAR, FACE, & NECK	88	48.7	90	48.7	64	35.8	78	42.9	82	43.1	46	23.9	51	27.0
EYE	26	14.4	30	16.2	19	10.6	25	13.7	23	12.1	7	3.6	18	9.5
Anophthalmia	2	1.1	1	0.5	1	0.6	0	0.0	0	0.0	1	0.5	0	0.0
Microphthalmia	3	1.7	3	1.6	3	1.7	3	1.6	4	2.1	1	0.5	4	2.1
Cataract	2	1.1	0	0.0	1	0.6	2	1.1	3	1.6	0	0.0	1	0.5
EAR	50	27.7	65	35.2	43	24.1	51	28.0	56	29.4	37	19.2	32	17.0
Absent auditory canal/ear	3	1.7	4	2.2	3	1.7	0	0.0	2	1.1	6	3.1	2	1.1
Microtia	6	3.3	7	3.8	4	2.2	9	4.9	11	5.8	2	1.0	7	3.7
CARDIAC & CIRCULATORY	311	172.1	364	197.0	347	194.2	359	197.4	377	198.1	295	153.4	344	182.3
Common truncus	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	1	0.5	2	1.1
Transposition of the great vessels	10	5.5	7	3.8	7	3.9	12	6.6	8	4.2	8	4.2	8	4.2
Tetralogy of Fallot	4	2.2	5	2.7	4	2.2	8	4.4	5	2.6	5	2.6	4	2.1
Single ventricle	3	1.7	2	1.1	0	0.0	4	2.2	0	0.0	0	0.0	0	0.0
Ventricular septal defect	83	45.9	66	35.7	77	43.1	63	34.6	76	39.9	72	37.4	62	32.8
Atrial septal defect	143	79.1	178	96.3	164	91.8	193	106.1	201	105.6	161	83.7	216	114.4
Endocardial cushion defect	1	0.6	7	3.8	1	0.6	5	2.7	3	1.6	2	1.0	4	2.1
Pulmonary valve atresia/stenosis	17	9.4	19	10.3	11	6.2	18	9.9	12	6.3	11	5.7	6	3.2
Tricuspid valve atresia/stenosis	1	0.6	6	3.2	2	1.1	5	2.7	2	1.1	2	1.0	1	0.5
Ebstein's anomaly	3	1.7	1	0.5	0	0.0	3	1.6	2	1.1	0	0.0	1	0.5
Aortic valve atresia/stenosis	1	0.6	2	1.1	1	0.6	2	1.1	1	0.5	1	0.5	1	0.5
Mitral valve atresia/stenosis	3	1.7	0	0.0	0	0.0	1	0.5	0	0.0	0	0.0	1	0.5
Hypoplastic left heart syndrome	4	2.2	3	1.6	1	0.6	1	0.5	0	0.0	3	1.6	2	1.1
Dextrocardia	2	1.1	2	1.1	0	0.0	6	3.3	1	0.5	0	0.0	0	0.0
Patent ductus arteriosus +	136	75.3	191	103.4	179	100.2	180	99.0	202	106.1	135	70.2	184	97.5
Coarctation of the aorta	2	1.1	3	1.6	4	2.2	6	3.3	9	4.7	5	2.6	3	1.6

*Total births=live births+fetal deaths **N=live births, fetal deaths, and terminations with birth defects ***Rate=(number of birth defects)/(total births)x10,000 +Birth weight >2,499 grams

TABLE 2. RATE OF BIRTH DEFECTS PER 10,000 TOTAL BIRTHS* BY YEAR, HAWAI'I, 1986-2005

Diagnosis	Total 1986-2005 N=394,569*		1986 N=19,819		1987 N=20,097		1988 N=20,541		1989 N=20,954		1990 N=22,288		1991 N=21,765	
	N**	Rate***	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Aortic stenosis	3	0.1	0	0.0	0	0.0	1	0.5	1	0.5	0	0.0	0	0.0
Pulmonary artery atresia/stenosis	736	18.7	11	5.6	15	7.5	13	6.3	7	3.3	17	7.6	22	10.1
Total anomalous pulmonary venous return	37	0.9	2	1.0	1	0.5	0	0.0	1	0.5	2	0.9	3	1.4
Partial anomalous pulmonary venous return	17	0.4	2	1.0	0	0.0	0	0.0	0	0.0	0	0.0	1	0.5
Persistent fetal circulation	214	5.4	31	15.6	38	18.9	21	10.2	24	11.5	17	7.6	18	8.3
RESPIRATORY	939	23.8	62	31.3	51	25.4	60	29.2	59	28.2	58	26.0	54	24.8
Choanal atresia/stenosis	50	1.3	4	2.0	3	1.5	3	1.5	4	1.9	7	3.1	1	0.5
Lung cysts	31	0.8	1	0.5	0	0.0	0	0.0	1	0.5	0	0.0	2	0.9
Hypoplastic lung	212	5.4	24	12.1	17	8.5	23	11.2	18	8.6	12	5.4	13	6.0
OROFACIAL & GASTROINTESTINAL	2,227	56.4	124	62.6	127	63.2	151	73.5	140	66.8	145	65.1	149	68.5
Oral clefts	735	18.6	35	17.7	45	22.4	47	22.9	33	15.7	40	17.9	42	19.3
Cleft palate (isolated)	275	7.0	11	5.6	14	7.0	21	10.2	6	2.9	20	9.0	14	6.4
Cleft lip (+/-cleft palate)	461	11.7	24	12.1	31	15.4	26	12.7	27	12.9	20	9.0	28	12.9
TE fistula/Esophageal atresia	78	2.0	9	4.5	3	1.5	7	3.4	8	3.8	6	2.7	2	0.9
Pyloric stenosis	287	7.3	13	6.6	16	8.0	20	9.7	30	14.3	19	8.5	16	7.4
Atresia/stenosis small intestine	108	2.7	2	1.0	4	2.0	4	1.9	7	3.3	7	3.1	5	2.3
Anal atresia/stenosis	184	4.7	10	5.0	8	4.0	11	5.4	11	5.2	13	5.8	13	6.0
Hirschsprung's disease	93	2.4	8	4.0	8	4.0	9	4.4	4	1.9	4	1.8	5	2.3
Malrotation	101	2.6	5	2.5	5	2.5	5	2.4	6	2.9	8	3.6	4	1.8
Biliary atresia	40	1.0	1	0.5	3	1.5	3	1.5	2	1.0	5	2.2	3	1.4
GENITAL & URINARY	4,094	103.8	216	109.0	234	116.4	236	114.9	232	110.7	257	115.3	239	109.8
Ovarian cyst	19	0.5	0	0.0	0	0.0	0	0.0	2	1.0	1	0.4	2	0.9
Undescended testicle +	1,169	29.6	86	43.4	87	43.3	101	49.2	77	36.7	90	40.4	77	35.4
Hypospadias/Epispadias/Chordee	1,258	31.9	43	21.7	69	34.3	53	25.8	56	26.7	62	27.8	59	27.1
Ambiguous genitalia	20	0.5	1	0.5	1	0.5	0	0.0	2	1.0	1	0.4	2	0.9
Absent/hypoplastic kidney	173	4.4	14	7.1	6	3.0	10	4.9	9	4.3	5	2.2	15	6.9
Cystic kidney	165	4.2	11	5.6	7	3.5	7	3.4	13	6.2	5	2.2	7	3.2
Hydronephrosis	515	13.1	12	6.1	15	7.5	13	6.3	27	12.9	30	13.5	31	14.2
Exstrophy bladder	9	0.2	1	0.5	0	0.0	0	0.0	1	0.5	0	0.0	0	0.0
Posterior urethral valves	23	0.6	4	2.0	0	0.0	1	0.5	2	1.0	1	0.4	2	0.9
LIMB & MUSCULOSKELETAL	4,928	124.9	316	159.4	320	159.2	333	162.1	331	158.0	361	162.0	298	136.9
Scoliosis	95	2.4	7	3.5	8	4.0	5	2.4	3	1.4	6	2.7	4	1.8
Hip dislocation	374	9.5	22	11.1	27	13.4	37	18.0	39	18.6	47	21.1	33	15.2

*Total births=live births+fetal deaths **N=live births, fetal deaths, and terminations with birth defects ***Rate=(number of birth defects)/(total births)x10,000 +Birth weight >2,499 grams

TABLE 2. RATE OF BIRTH DEFECTS PER 10,000 TOTAL BIRTHS* BY YEAR, HAWAI'I, 1986-2005

Diagnosis	1992		1993		1994		1995		1996		1997		1998	
	N=21,504		N=21,073		N=20,814		N=19,750		N=19,386		N=18,198		N=19,644	
	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Aortic stenosis	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Pulmonary artery atresia/stenosis	19	8.8	33	15.7	48	23.1	57	28.9	61	31.5	66	36.3	44	23.6
Total anomalous pulmonary venous return	2	0.9	3	1.4	0	0.0	4	2.0	4	2.1	1	0.5	1	0.5
Partial anomalous pulmonary venous return	1	0.5	0	0.0	1	0.5	3	1.5	2	1.0	0	0.0	1	0.5
Persistent fetal circulation	10	4.7	8	3.8	12	5.8	4	2.0	2	1.0	0	0.0	2	1.1
RESPIRATORY	55	25.6	50	23.7	57	27.4	69	34.9	75	38.7	45	24.7	48	25.7
Choanal atresia/stenosis	3	1.4	1	0.5	5	2.4	3	1.5	4	2.1	0	0.0	3	1.6
Lung cysts	4	1.9	0	0.0	1	0.5	1	0.5	3	1.5	2	1.1	0	0.0
Hypoplastic lung	7	3.3	10	4.7	6	2.9	13	6.6	9	4.6	10	5.5	4	2.1
OROFACIAL & GASTROINTESTINAL	131	60.9	110	52.2	121	58.1	127	64.3	158	81.5	105	57.7	96	51.5
Oral clefts	49	22.8	41	19.5	33	15.9	45	22.8	43	22.2	46	25.3	41	22.0
Cleft palate (isolated)	19	8.8	19	9.0	12	5.8	17	8.6	15	7.7	16	8.8	17	9.1
Cleft lip (+/-cleft palate)	30	14.0	22	10.4	22	10.6	28	14.2	28	14.4	30	16.5	24	12.9
TE fistula/Esophageal atresia	1	0.5	2	0.9	7	3.4	1	0.5	8	4.1	2	1.1	2	1.1
Pyloric stenosis	15	7.0	18	8.5	15	7.2	17	8.6	22	11.3	12	6.6	6	3.2
Atresia/stenosis small intestine	5	2.3	3	1.4	6	2.9	6	3.0	12	6.2	6	3.3	8	4.3
Anal atresia/stenosis	8	3.7	9	4.3	10	4.8	7	3.5	16	8.3	8	4.4	5	2.7
Hirschsprung's disease	5	2.3	4	1.9	5	2.4	5	2.5	4	2.1	3	1.6	7	3.8
Malrotation	6	2.8	4	1.9	7	3.4	11	5.6	11	5.7	5	2.7	5	2.7
Biliary atresia	4	1.9	1	0.5	1	0.5	1	0.5	4	2.1	1	0.5	0	0.0
GENITAL & URINARY	239	111.1	234	111.0	210	100.9	245	124.1	279	143.9	190	104.4	185	99.2
Ovarian cyst	1	0.5	2	0.9	1	0.5	1	0.5	1	0.5	3	1.6	0	0.0
Undescended testicle +	99	46.0	93	44.1	80	38.4	70	35.4	90	46.4	53	29.1	64	34.3
Hypospadias/Epispadias/Chordee	58	27.0	60	28.5	58	27.9	74	37.5	71	36.6	53	29.1	58	31.1
Ambiguous genitalia	0	0.0	0	0.0	0	0.0	1	0.5	3	1.5	0	0.0	1	0.5
Absent/hypoplastic kidney	9	4.2	6	2.8	11	5.3	10	5.1	16	8.3	9	4.9	7	3.8
Cystic kidney	14	6.5	6	2.8	6	2.9	13	6.6	11	5.7	10	5.5	6	3.2
Hydronephrosis	27	12.6	26	12.3	18	8.6	27	13.7	25	12.9	23	12.6	27	14.5
Exstrophy bladder	2	0.9	0	0.0	0	0.0	0	0.0	1	0.5	0	0.0	2	1.1
Posterior urethral valves	0	0.0	1	0.5	0	0.0	5	2.5	2	1.0	0	0.0	0	0.0
LIMB & MUSCULOSKELETAL	289	134.4	277	131.4	284	136.4	264	133.7	269	138.8	232	127.5	202	108.3
Scoliosis	5	2.3	2	0.9	10	4.8	3	1.5	6	3.1	8	4.4	3	1.6
Hip dislocation	25	11.6	29	13.8	15	7.2	13	6.6	6	3.1	11	6.0	10	5.4

*Total births=live births+fetal deaths **N=live births, fetal deaths, and terminations with birth defects ***Rate=(number of birth defects)/(total births)x10,000 +Birth weight >2,499 grams

TABLE 2. RATE OF BIRTH DEFECTS PER 10,000 TOTAL BIRTHS* BY YEAR, HAWAI'I, 1986-2005

Diagnosis	1999 N=18,067		2000 N=18,478		2001 N=17867		2002 N=18,182		2003 N=19,033		2004 N=19,234		2005 N=18,875	
	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Aortic stenosis	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	1	0.5
Pulmonary artery atresia/stenosis	33	18.3	49	26.5	44	24.6	40	22.0	62	32.6	48	25.0	47	24.9
Total anomalous pulmonary venous return	0	0.0	3	1.6	1	0.6	3	1.6	1	0.5	2	1.0	3	1.6
Partial anomalous pulmonary venous return	2	1.1	2	1.1	2	1.1	0	0.0	0	0.0	0	0.0	0	0.0
Persistent fetal circulation	2	1.1	1	0.5	0	0.0	2	1.1	9	4.7	8	4.2	5	2.6
RESPIRATORY	37	20.5	38	20.6	36	20.1	28	15.4	27	14.2	19	9.9	11	5.8
Choanal atresia/stenosis	3	1.7	1	0.5	1	0.6	3	1.6	1	0.5	0	0.0	0	0.0
Lung cysts	1	0.6	4	2.2	2	1.1	2	1.1	4	2.1	2	1.0	1	0.5
Hypoplastic lung	8	4.4	8	4.3	12	6.7	4	2.2	11	5.8	3	1.6	0	0.0
OROFACIAL & GASTROINTESTINAL	74	41.0	99	53.6	68	38.1	72	39.6	83	43.6	63	32.8	84	44.5
Oral clefts	27	14.9	35	18.9	23	12.9	27	14.8	26	13.7	21	10.9	36	19.1
Cleft palate (isolated)	6	3.3	14	7.6	6	3.4	12	6.6	10	5.3	10	5.2	16	8.5
Cleft lip (+/-cleft palate)	21	11.6	21	11.4	17	9.5	15	8.2	16	8.4	11	5.7	20	10.6
TE fistula/Esophageal atresia	4	2.2	2	1.1	5	2.8	1	0.5	1	0.5	4	2.1	3	1.6
Pyloric stenosis	10	5.5	9	4.9	8	4.5	14	7.7	12	6.3	7	3.6	8	4.2
Atresia/stenosis small intestine	2	1.1	10	5.4	1	0.6	3	1.6	5	2.6	2	1.0	10	5.3
Anal atresia/stenosis	9	5.0	14	7.6	7	3.9	5	2.7	7	3.7	9	4.7	4	2.1
Hirschsprung's disease	3	1.7	4	2.2	2	1.1	1	0.5	3	1.6	4	2.1	5	2.6
Malrotation	5	2.8	3	1.6	4	2.2	1	0.5	2	1.1	1	0.5	3	1.6
Biliary atresia	2	1.1	2	1.1	0	0.0	1	0.5	2	1.1	2	1.0	2	1.1
GENITAL & URINARY	144	79.7	146	79.0	175	97.9	157	86.3	173	90.9	133	69.1	170	90.1
Ovarian cyst	0	0.0	2	1.1	1	0.6	0	0.0	1	0.5	1	0.5	0	0.0
Undescended testicle +	16	8.9	12	6.5	14	7.8	12	6.6	17	8.9	17	8.8	14	7.4
Hypospadias/Epispadias/Chordee	67	37.1	68	36.8	84	47.0	76	41.8	67	35.2	53	27.6	69	36.6
Ambiguous genitalia	0	0.0	1	0.5	2	1.1	0	0.0	0	0.0	4	2.1	1	0.5
Absent/hypoplastic kidney	9	5.0	7	3.8	10	5.6	2	1.1	12	6.3	4	2.1	2	1.1
Cystic kidney	11	6.1	8	4.3	8	4.5	4	2.2	7	3.7	4	2.1	7	3.7
Hydronephrosis	19	10.5	16	8.7	33	18.5	26	14.3	40	21.0	33	17.2	47	24.9
Exstrophy bladder	2	1.1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Posterior urethral valves	1	0.6	1	0.5	0	0.0	1	0.5	1	0.5	1	0.5	0	0.0
LIMB & MUSCULOSKELETAL	193	106.8	172	93.1	167	93.5	160	88.0	170	89.3	133	69.1	157	83.2
Scoliosis	6	3.3	5	2.7	1	0.6	3	1.6	5	2.6	4	2.1	1	0.5
Hip dislocation	11	6.1	11	6.0	8	4.5	5	2.7	11	5.8	3	1.6	11	5.8

*Total births=live births+fetal deaths **N=live births, fetal deaths, and terminations with birth defects ***Rate=(number of birth defects)/(total births)x10,000 +Birth weight >2,499 grams

TABLE 2. RATE OF BIRTH DEFECTS PER 10,000 TOTAL BIRTHS* BY YEAR, HAWAI'I, 1986-2005

Diagnosis	Total 1986-2005 N=394,569*		1986 N=19,819		1987 N=20,097		1988 N=20,541		1989 N=20,954		1990 N=22,288		1991 N=21,765	
	N**	Rate***	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Clubfoot	1,206	30.6	110	55.5	88	43.8	94	45.8	63	30.1	76	34.1	60	27.6
Polydactyly	642	16.3	25	12.6	32	15.9	38	18.5	49	23.4	48	21.5	28	12.9
Syndactyly	298	7.6	22	11.1	13	6.5	11	5.4	15	7.2	25	11.2	20	9.2
Limb reduction deformity - upper	129	3.3	9	4.5	5	2.5	7	3.4	8	3.8	8	3.6	8	3.7
Limb reduction deformity - lower	54	1.4	1	0.5	1	0.5	4	1.9	2	1.0	5	2.2	3	1.4
Craniosynostosis	181	4.6	15	7.6	12	6.0	12	5.8	17	8.1	16	7.2	10	4.6
Crouzon's disease	2	0.1	0	0.0	0	0.0	0	0.0	0	0.0	1	0.4	0	0.0
Apert syndrome	11	0.3	1	0.5	0	0.0	0	0.0	1	0.5	1	0.4	0	0.0
Goldenhar syndrome	21	0.5	1	0.5	1	0.5	2	1.0	2	1.0	1	0.4	0	0.0
Spina bifida occulta	13	0.3	0	0.0	0	0.0	3	1.5	1	0.5	0	0.0	1	0.5
Dwarfism	103	2.6	9	4.5	5	2.5	5	2.4	5	2.4	7	3.1	2	0.9
Osteogenesis imperfecta	15	0.4	0	0.0	0	0.0	1	0.5	2	1.0	1	0.4	1	0.5
Diaphragmatic hernia	88	2.2	7	3.5	5	2.5	7	3.4	10	4.8	6	2.7	5	2.3
Abdominal wall defects	322	8.2	15	7.6	9	4.5	14	6.8	17	8.1	14	6.3	16	7.4
Omphalocele	100	2.5	6	3.0	2	1.0	4	1.9	8	3.8	3	1.3	6	2.8
Gastroschisis	132	3.3	6	3.0	2	1.0	3	1.5	6	2.9	4	1.8	6	2.8
Prune-belly syndrome	10	0.3	0	0.0	0	0.0	1	0.5	0	0.0	0	0.0	0	0.0
Poland syndrome	6	0.2	0	0.0	0	0.0	0	0.0	1	0.5	1	0.4	0	0.0
SKIN & INTEGUMENT	2,296	58.2	99	50.0	161	80.1	158	76.9	193	92.1	194	87.0	162	74.4
CHROMOSOME & SYNDROME	2,087	52.9	78	39.4	89	44.3	108	52.6	106	50.6	118	52.9	148	68.0
Down syndrome (trisomy 21)	566	14.3	29	14.6	26	12.9	39	19.0	36	17.2	28	12.6	39	17.9
Patau syndrome (trisomy 13)	75	1.9	1	0.5	0	0.0	2	1.0	3	1.4	6	2.7	5	2.3
Edwards syndrome (trisomy 18)	168	4.3	3	1.5	6	3.0	3	1.5	8	3.8	14	6.3	12	5.5
Cri du Chat syndrome	8	0.2	0	0.0	0	0.0	1	0.5	0	0.0	0	0.0	1	0.5
Wolff-Hirschorn syndrome	8	0.2	0	0.0	0	0.0	0	0.0	0	0.0	1	0.4	0	0.0
Autosomal anomalies in normal infant	142	3.6	8	4.0	6	3.0	8	3.9	6	2.9	9	4.0	13	6.0
Triploidy	46	1.2	3	1.5	1	0.5	1	0.5	1	0.5	5	2.2	2	0.9
Turner syndrome (45,X)	149	3.8	3	1.5	10	5.0	3	1.5	9	4.3	7	3.1	13	6.0
Klinefelter syndrome (47,XXY)	55	1.4	1	0.5	3	1.5	2	1.0	4	1.9	3	1.3	4	1.8
Absent spleen/Asplenia	13	0.3	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Accessory spleen	42	1.1	5	2.5	1	0.5	4	1.9	2	1.0	0	0.0	4	1.8
Situs inversus	43	1.1	2	1.0	2	1.0	1	0.5	3	1.4	0	0.0	2	0.9
Conjoined twins	13	0.3	0	0.0	0	0.0	0	0.0	2	1.0	0	0.0	0	0.0

*Total births=live births+fetal deaths **N=live births, fetal deaths, and terminations with birth defects ***Rate=(number of birth defects)/(total births)x10,000 +Birth weight >2,499 grams

TABLE 2. RATE OF BIRTH DEFECTS PER 10,000 TOTAL BIRTHS* BY YEAR, HAWAI‘I, 1986-2005

Diagnosis	1992 N=21,504		1993 N=21,073		1994 N=20,814		1995 N=19,750		1996 N=19,386		1997 N=18,198		1998 N=19,644	
	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Clubfoot	64	29.8	61	28.9	63	30.3	45	22.8	72	37.1	56	30.8	40	21.5
Polydactyly	37	17.2	34	16.1	33	15.9	37	18.7	41	21.1	37	20.3	31	16.6
Syndactyly	25	11.6	18	8.5	26	12.5	14	7.1	12	6.2	24	13.2	12	6.4
Limb reduction deformity - upper	9	4.2	8	3.8	8	3.8	3	1.5	8	4.1	5	2.7	11	5.9
Limb reduction deformity - lower	4	1.9	4	1.9	1	0.5	3	1.5	1	0.5	2	1.1	6	3.2
Craniosynostosis	9	4.2	21	10.0	24	11.5	16	8.1	6	3.1	2	1.1	4	2.1
Crouzon’s disease	1	0.5	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Apert syndrome	0	0.0	1	0.5	2	1.0	1	0.5	0	0.0	1	0.5	0	0.0
Goldenhar syndrome	1	0.5	0	0.0	1	0.5	2	1.0	3	1.5	1	0.5	0	0.0
Spina bifida occulta	0	0.0	0	0.0	1	0.5	1	0.5	0	0.0	2	1.1	1	0.5
Dwarfism	8	3.7	8	3.8	7	3.4	8	4.1	8	4.1	4	2.2	6	3.2
Osteogenesis imperfecta	1	0.5	1	0.5	2	1.0	0	0.0	1	0.5	1	0.5	2	1.1
Diaphragmatic hernia	5	2.3	4	1.9	2	1.0	5	2.5	4	2.1	5	2.7	3	1.6
Abdominal wall defects	17	7.9	14	6.6	24	11.5	26	13.2	24	12.4	24	13.2	21	11.3
Omphalocele	6	2.8	3	1.4	10	4.8	5	2.5	8	4.1	7	3.8	8	4.3
Gastroschisis	5	2.3	9	4.3	9	4.3	10	5.1	7	3.6	7	3.8	10	5.4
Prune-belly syndrome	1	0.5	1	0.5	0	0.0	0	0.0	2	1.0	1	0.5	0	0.0
Poland syndrome	1	0.5	0	0.0	0	0.0	1	0.5	0	0.0	0	0.0	1	0.5
SKIN & INTEGUMENT	58	27.0	63	29.9	73	35.1	103	52.2	181	93.4	175	96.2	93	49.9
CHROMOSOME & SYNDROME	132	61.4	115	54.6	141	67.7	128	64.8	153	78.9	127	69.8	91	48.8
Down syndrome (trisomy 21)	32	14.9	27	12.8	32	15.4	23	11.6	37	19.1	31	17.0	30	16.1
Patau syndrome (trisomy 13)	8	3.7	5	2.4	5	2.4	3	1.5	5	2.6	8	4.4	2	1.1
Edwards syndrome (trisomy 18)	8	3.7	10	4.7	13	6.2	14	7.1	18	9.3	10	5.5	5	2.7
Cri du Chat syndrome	0	0.0	2	0.9	1	0.5	1	0.5	0	0.0	1	0.5	0	0.0
Wolff-Hirschorn syndrome	1	0.5	0	0.0	2	1.0	2	1.0	0	0.0	1	0.5	0	0.0
Autosomal anomalies in normal infant	11	5.1	6	2.8	19	9.1	13	6.6	10	5.2	8	4.4	4	2.1
Triploidy	5	2.3	4	1.9	4	1.9	3	1.5	5	2.6	2	1.1	0	0.0
Turner syndrome (45,X)	12	5.6	4	1.9	8	3.8	11	5.6	16	8.3	6	3.3	5	2.7
Klinefelter syndrome (47,XXY)	2	0.9	4	1.9	2	1.0	1	0.5	6	3.1	4	2.2	3	1.6
Absent spleen/Asplenia	0	0.0	1	0.5	0	0.0	3	1.5	2	1.0	2	1.1	1	0.5
Accessory spleen	2	0.9	2	0.9	3	1.4	5	2.5	3	1.5	1	0.5	1	0.5
Situs inversus	4	1.9	1	0.5	2	1.0	3	1.5	4	2.1	4	2.2	2	1.1
Conjoined twins	0	0.0	0	0.0	0	0.0	2	1.0	2	1.0	4	2.2	0	0.0

*Total births=live births+fetal deaths **N=live births, fetal deaths, and terminations with birth defects ***Rate=(number of birth defects)/(total births)x10,000 +Birth weight >2,499 grams

TABLE 2. RATE OF BIRTH DEFECTS PER 10,000 TOTAL BIRTHS* BY YEAR, HAWAI‘I, 1986-2005

Diagnosis	1999 N=18,067		2000 N=18,478		2001 N=17867		2002 N=18,182		2003 N=19,033		2004 N=19,234		2005 N=18,875	
	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Clubfoot	57	31.5	45	24.4	37	20.7	40	22.0	55	28.9	36	18.7	44	23.3
Polydactyly	26	14.4	23	12.4	31	17.4	25	13.7	26	13.7	24	12.5	17	9.0
Syndactyly	12	6.6	13	7.0	7	3.9	8	4.4	8	4.2	6	3.1	7	3.7
Limb reduction deformity - upper	4	2.2	7	3.8	3	1.7	5	2.7	3	1.6	5	2.6	5	2.6
Limb reduction deformity - lower	2	1.1	4	2.2	3	1.7	2	1.1	3	1.6	2	1.0	1	0.5
Craniosynostosis	2	1.1	2	1.1	2	1.1	4	2.2	1	0.5	2	1.0	4	2.1
Crouzon’s disease	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Apert syndrome	1	0.6	1	0.5	1	0.6	0	0.0	0	0.0	0	0.0	0	0.0
Goldenhar syndrome	0	0.0	2	1.1	0	0.0	2	1.1	0	0.0	1	0.5	1	0.5
Spina bifida occulta	1	0.6	0	0.0	1	0.6	0	0.0	0	0.0	0	0.0	1	0.5
Dwarfism	3	1.7	1	0.5	5	2.8	3	1.6	2	1.1	4	2.1	3	1.6
Osteogenesis imperfecta	0	0.0	1	0.5	0	0.0	1	0.5	0	0.0	0	0.0	0	0.0
Diaphragmatic hernia	7	3.9	3	1.6	2	1.1	1	0.5	4	2.1	0	0.0	3	1.6
Abdominal wall defects	14	7.7	12	6.5	11	6.2	15	8.2	13	6.8	8	4.2	14	7.4
Omphalocele	4	2.2	6	3.2	1	0.6	3	1.6	4	2.1	2	1.0	4	2.1
Gastroschisis	6	3.3	3	1.6	8	4.5	9	4.9	8	4.2	5	2.6	9	4.8
Prune-belly syndrome	0	0.0	1	0.5	2	1.1	0	0.0	0	0.0	1	0.5	0	0.0
Poland syndrome	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	1	0.5	0	0.0
SKIN & INTEGUMENT	79	43.7	105	56.8	87	48.7	81	44.5	90	47.3	63	32.8	78	41.3
CHROMOSOME & SYNDROME	110	60.9	85	46.0	63	35.3	71	39.0	61	32.0	89	46.3	74	39.2
Down syndrome (trisomy 21)	25	13.8	26	14.1	20	11.2	22	12.1	18	9.5	30	15.6	16	8.5
Patau syndrome (trisomy 13)	6	3.3	4	2.2	2	1.1	1	0.5	1	0.5	4	2.1	4	2.1
Edwards syndrome (trisomy 18)	14	7.7	8	4.3	3	1.7	7	3.8	5	2.6	4	2.1	3	1.6
Cri du Chat syndrome	1	0.6	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Wolff-Hirschorn syndrome	1	0.6	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Autosomal anomalies in normal infant	5	2.8	2	1.1	6	3.4	0	0.0	3	1.6	3	1.6	2	1.1
Triploidy	2	1.1	2	1.1	1	0.6	2	1.1	1	0.5	1	0.5	1	0.5
Turner syndrome (45,X)	13	7.2	8	4.3	4	2.2	2	1.1	6	3.2	6	3.1	3	1.6
Klinefelter syndrome (47,XXY)	4	2.2	3	1.6	2	1.1	0	0.0	3	1.6	3	1.6	1	0.5
Absent spleen/Asplenia	1	0.6	0	0.0	0	0.0	1	0.5	0	0.0	0	0.0	2	1.1
Accessory spleen	2	1.1	3	1.6	1	0.6	2	1.1	0	0.0	0	0.0	1	0.5
Situs inversus	1	0.6	2	1.1	1	0.6	2	1.1	1	0.5	2	1.0	4	2.1
Conjoined twins	0	0.0	0	0.0	2	1.1	0	0.0	0	0.0	0	0.0	1	0.5

*Total births=live births+fetal deaths **N=live births, fetal deaths, and terminations with birth defects ***Rate=(number of birth defects)/(total births)x10,000 +Birth weight >2,499 grams

TABLE 2. RATE OF BIRTH DEFECTS PER 10,000 TOTAL BIRTHS* BY YEAR, HAWAI'I, 1986-2005

Diagnosis	Total 1986-2005 N=394,569*		1986 N=19,819		1987 N=20,097		1988 N=20,541		1989 N=20,954		1990 N=22,288		1991 N=21,765	
	N**	Rate***	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Tuberous sclerosis	15	0.4	0	0.0	0	0.0	1	0.5	0	0.0	2	0.9	2	0.9
Amniotic band syndrome	82	2.1	8	4.0	4	2.0	4	1.9	10	4.8	4	1.8	6	2.8
DiGeorge syndrome	25	0.6	0	0.0	1	0.5	1	0.5	2	1.0	2	0.9	0	0.0
Pierre-Robin sequence	36	0.9	2	1.0	4	2.0	3	1.5	0	0.0	1	0.4	1	0.5
Werdnig-Hoffman syndrome	16	0.4	0	0.0	1	0.5	0	0.0	1	0.5	3	1.3	0	0.0
Wolff-Parkinson-White syndrome	20	0.5	0	0.0	1	0.5	1	0.5	1	0.5	1	0.4	1	0.5
NEOPLASMS	375	9.5	20	10.1	19	9.5	25	12.2	24	11.5	20	9.0	23	10.6
TOTAL BIRTH DEFECTS CASES	17,311	438.7	825	416.3	1,027	511.0	979	476.6	943	450.0	983	441.0	903	414.9
FETAL ALCOHOL SYNDROME	90	2.3	4	2.0	5	2.5	5	2.4	10	4.8	10	4.5	10	4.6
BIRTH DEFECT CASES WITH:														
CONGENITAL INFECTION	269	6.8	17	8.6	28	13.9	22	10.7	27	12.9	17	7.6	16	7.4
MATERNAL ALCOHOL USE	1,528	38.7	69	34.8	80	39.8	103	50.1	132	63.0	114	51.1	109	50.1
MATERNAL TOBACCO USE	5,004	126.8	138	69.6	195	97.0	206	100.3	280	133.6	265	118.9	256	117.6
MATERNAL ILLICIT DRUG USE	3,194	80.9	24	12.1	30	14.9	43	20.9	140	66.8	118	52.9	110	50.5
Marijuana	1,012	25.6	10	5.0	17	8.5	20	9.7	54	25.8	39	17.5	36	16.5
Cocaine	591	15.0	15	7.6	18	9.0	25	12.2	57	27.2	52	23.3	37	17.0
Methamphetamine/Amphetamine	2,132	54.0	2	1.0	1	0.5	7	3.4	63	30.1	45	20.2	58	26.6

*Total births=live births+fetal deaths **N=live births, fetal deaths, and terminations with birth defects ***Rate=(number of birth defects)/(total births)x10,000 +Birth weight >2,499 grams

TABLE 2. RATE OF BIRTH DEFECTS PER 10,000 TOTAL BIRTHS* BY YEAR, HAWAI'I, 1986-2005

Diagnosis	1992 N=21,504		1993 N=21,073		1994 N=20,814		1995 N=19,750		1996 N=19,386		1997 N=18,198		1998 N=19,644	
	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Tuberous sclerosis	0	0.0	1	0.5	1	0.5	0	0.0	2	1.0	1	0.5	1	0.5
Amniotic band syndrome	9	4.2	5	2.4	4	1.9	2	1.0	1	0.5	4	2.2	6	3.2
DiGeorge syndrome	1	0.5	1	0.5	2	1.0	2	1.0	4	2.1	2	1.1	3	1.6
Pierre-Robin sequence	2	0.9	4	1.9	1	0.5	2	1.0	1	0.5	2	1.1	2	1.1
Werdnig-Hoffman syndrome	0	0.0	1	0.5	1	0.5	2	1.0	0	0.0	1	0.5	0	0.0
Wolff-Parkinson-White syndrome	1	0.5	2	0.9	1	0.5	1	0.5	0	0.0	1	0.5	1	0.5
NEOPLASMS	32	14.9	16	7.6	29	13.9	20	10.1	32	16.5	20	11.0	10	5.4
TOTAL BIRTH DEFECTS CASES	868	403.6	932	442.3	1,007	483.8	1,024	518.5	1,135	585.5	925	508.3	848	454.8
FETAL ALCOHOL SYNDROME	5	2.3	5	2.4	6	2.9	14	7.1	5	2.6	4	2.2	2	1.1
BIRTH DEFECT CASES WITH:														
CONGENITAL INFECTION	10	4.7	12	5.7	19	9.1	17	8.6	19	9.8	16	8.8	9	4.8
MATERNAL ALCOHOL USE	75	34.9	82	38.9	92	44.2	105	53.2	113	58.3	84	46.2	61	32.7
MATERNAL TOBACCO USE	220	102.3	240	113.9	277	133.1	315	159.5	326	168.2	330	181.3	289	155.0
MATERNAL ILLICIT DRUG USE	116	53.9	164	77.8	194	93.2	215	108.9	190	98.0	235	129.1	204	109.4
Marijuana	20	9.3	39	18.5	41	19.7	64	32.4	64	33.0	66	36.3	75	40.2
Cocaine	36	16.7	35	16.6	45	21.6	42	21.3	46	23.7	50	27.5	23	12.3
Methamphetamine/Amphetamine	69	32.1	113	53.6	126	60.5	151	76.5	113	58.3	149	81.9	140	75.1

*Total births=live births+fetal deaths **N=live births, fetal deaths, and terminations with birth defects ***Rate=(number of birth defects)/(total births)x10,000 +Birth weight >2,499 grams

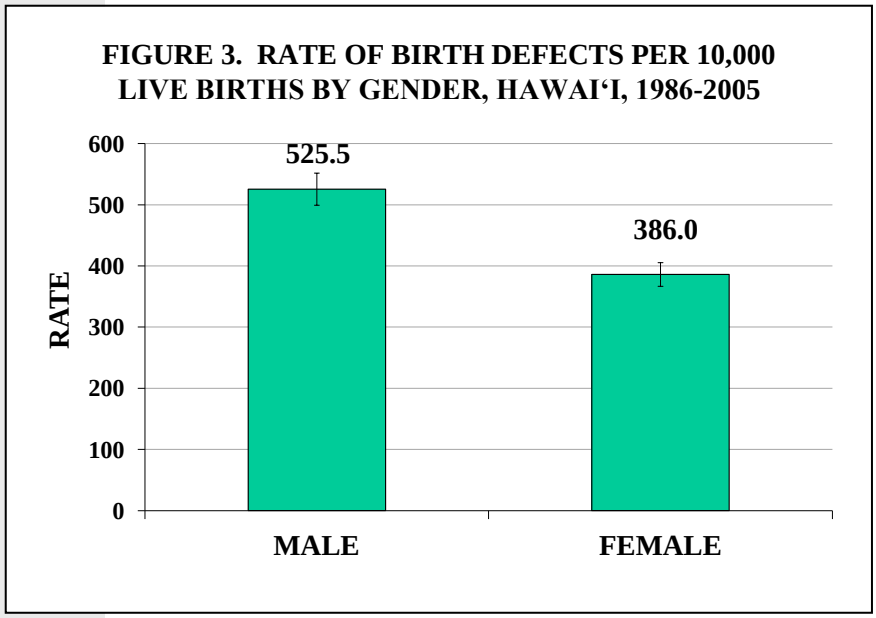
TABLE 2. RATE OF BIRTH DEFECTS PER 10,000 TOTAL BIRTHS* BY YEAR, HAWAI'I, 1986-2005

Diagnosis	1999 N=18,067		2000 N=18,478		2001 N=17867		2002 N=18,182		2003 N=19,033		2004 N=19,234		2005 N=18,875	
	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Tuberous sclerosis	0	0.0	0	0.0	1	0.6	1	0.5	0	0.0	1	0.5	1	0.5
Amniotic band syndrome	2	1.1	5	2.7	1	0.6	2	1.1	0	0.0	2	1.0	3	1.6
DiGeorge syndrome	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	3	1.6	1	0.5
Pierre-Robin sequence	0	0.0	3	1.6	1	0.6	2	1.1	0	0.0	2	1.0	3	1.6
Werdnig-Hoffman syndrome	1	0.6	0	0.0	0	0.0	1	0.5	2	1.1	1	0.5	1	0.5
Wolff-Parkinson-White syndrome	2	1.1	1	0.5	2	1.1	1	0.5	1	0.5	0	0.0	1	0.5
NEOPLASMS	17	9.4	10	5.4	11	6.2	14	7.7	14	7.4	9	4.7	10	5.3
TOTAL BIRTH DEFECTS CASES	693	383.6	729	394.5	722	404.1	699	384.4	717	376.7	637	331.2	715	378.8
FETAL ALCOHOL SYNDROME	1	0.6	0	0.0	1	0.6	2	1.1	0	0.0	1	0.5	0	0.0
BIRTH DEFECT CASES WITH:														
CONGENITAL INFECTION	12	6.6	6	3.2	5	2.8	10	5.5	5	2.6	0	0.0	2	1.1
MATERNAL ALCOHOL USE	55	30.4	51	27.6	46	25.7	52	28.6	39	20.5	38	19.8	28	14.8
MATERNAL TOBACCO USE	225	124.5	232	125.6	263	147.2	261	143.5	251	131.9	209	108.7	226	119.7
MATERNAL ILLICIT DRUG USE	170	94.1	219	118.5	234	131.0	208	114.4	206	108.2	194	100.9	180	95.4
Marijuana	62	34.3	89	48.2	77	43.1	67	36.8	80	42.0	44	22.9	48	25.4
Cocaine	18	10.0	27	14.6	22	12.3	18	9.9	8	4.2	9	4.7	8	4.2
Methamphetamine/Amphetamine	120	66.4	157	85.0	170	95.1	170	93.5	165	86.7	169	87.9	144	76.3

*Total births=live births+fetal deaths **N=live births, fetal deaths, and terminations with birth defects ***Rate=(number of birth defects)/(total births)x10,000 +Birth weight >2,499 grams

Birth Defect Rates by Gender

The overall birth defect rate for males (525.5 per 10,000 live births) was 36% higher than that for females (386.0), for 1986-2005 (Figure 3).



There were also differences among broad categories and specific birth defects by gender (Table 3).

TABLE 3. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY GENDER, HAWAI'I, 1986-2005

Diagnosis	Male		Female	
	N	Rate*	N	Rate
BRAIN & NERVOUS SYSTEM	741	38.8	730	40.7
Neural tube defects	131	6.9	169	9.4
Anencephaly	47	2.5	48	2.7
Spina bifida	60	3.1	92	5.1
Encephalocele	31	1.6	32	1.8
Microcephaly	161	8.4	207	11.5
Holoprosencephaly	21	1.1	25	1.4
Hydrocephaly (w/o spina bifida)	214	11.2	179	10.0
EYE, EAR, FACE, & NECK	1,042	54.6	837	46.6
EYE	348	18.2	358	19.9
Anophthalmia	6	0.3	7	0.4
Microphthalmia	52	2.7	56	3.1
Cataract	21	1.1	24	1.3
EAR	766	40.1	562	31.3
Absent auditory canal/ear	24	1.3	21	1.2
Microtia	85	4.5	45	2.5
CARDIAC & CIRCULATORY	3,757	196.8	3,306	184.1
Common truncus	16	0.8	11	0.6
Transposition of the great vessels	97	5.1	63	3.5
Tetralogy of Fallot	69	3.6	71	4.0
Single ventricle	12	0.6	15	0.8
Ventricular septal defect	858	45.0	895	49.8
Atrial septal defect	1,455	76.2	1,277	71.1
Endocardial cushion defect	36	1.9	50	2.8
Pulmonary valve atresia/stenosis	186	9.7	159	8.9
Tricuspid valve atresia/stenosis	27	1.4	30	1.7
Ebstein's anomaly	8	0.4	10	0.6
Aortic valve atresia/stenosis	36	1.9	16	0.9
Mitral valve atresia/stenosis	22	1.2	16	0.9
Hypoplastic left heart syndrome	32	1.7	25	1.4
Dextrocardia	25	1.3	28	1.6
Patent ductus arteriosus +	1,621	84.9	1,402	78.1
Coarctation of the aorta	65	3.4	40	2.2
Aortic stenosis	1	0.1	2	0.1
Pulmonary artery atresia/stenosis	393	20.6	338	18.8
Total anomalous pulmonary venous return	9	0.5	28	1.6
Partial anomalous pulmonary venous return	7	0.4	10	0.6
Persistent fetal circulation	125	6.5	89	5.0
RESPIRATORY	526	27.6	412	22.9
Choanal atresia/stenosis	29	1.5	21	1.2
Lung cysts	23	1.2	8	0.4
Hypoplastic lung	116	6.1	96	5.3
OROFACIAL & GASTROINTESTINAL	1,290	67.6	920	51.2
Oral clefts	405	21.2	321	17.9
Cleft palate (isolated)	118	6.2	154	8.6

* Rate = (number of birth defects)/(number of live births)x10,000.
+ Birth weight >2,499 grams.

TABLE 3. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY GENDER, HAWAI‘I, 1986-2005

Diagnosis	Male		Female	
	N	Rate*	N	Rate
Cleft lip (+/-cleft palate)	287	15.0	168	9.4
TE fistula/Esophageal atresia	41	2.1	37	2.1
Pyloric stenosis	235	12.3	49	2.7
Atresia/stenosis small intestine	62	3.2	45	2.5
Anal atresia/stenosis	106	5.6	76	4.2
Hirschsprung’s disease	66	3.5	27	1.5
Malrotation	53	2.8	48	2.7
Biliary atresia	20	1.0	19	1.1
GENITAL & URINARY	3,469	181.7	606	33.7
Ovarian cyst	0	0.0	19	1.1
Undescended testicle +	1,169	61.2	0	0.0
Hypospadias/Epispadias/Chordee	1,254	65.7	4	0.2
Ambiguous genitalia	7	0.4	12	0.7
Absent/hypoplastic kidney	97	5.1	74	4.1
Cystic kidney	82	4.3	81	4.5
Hydronephrosis	363	19.0	150	8.4
Exstrophy bladder	4	0.2	5	0.3
Posterior urethral valves	20	1.0	1	0.1
LIMB & MUSCULOSKELETAL	2,518	131.9	2,375	132.3
Scoliosis	48	2.5	42	2.3
Hip dislocation	100	5.2	274	15.3
Clubfoot	647	33.9	557	31.0
Polydactyly	365	19.1	273	15.2
Syndactyly	181	9.5	115	6.4
Limb reduction deformity - upper	84	4.4	45	2.5
Limb reduction deformity - lower	31	1.6	22	1.2
Craniosynostosis	98	5.1	83	4.6
Crouzon’s disease	0	0.0	2	0.1
Apert syndrome	4	0.2	7	0.4
Goldenhar syndrome	12	0.6	9	0.5
Spina bifida occulta	7	0.4	5	0.3
Dwarfism	50	2.6	52	2.9
Osteogenesis imperfecta	6	0.3	9	0.5
Diaphragmatic hernia	47	2.5	39	2.2
Abdominal wall defects	176	9.2	132	7.4
Omphalocele	51	2.7	41	2.3
Gastroschisis	75	3.9	55	3.1
Prune-belly syndrome	8	0.4	2	0.1
Poland syndrome	5	0.3	1	0.1
SKIN & INTEGUMENT	1,329	69.6	965	53.7
CHROMOSOME & SYNDROME	1,005	52.7	1,021	56.9
Down syndrome (trisomy 21)	293	15.4	265	14.8
Patau syndrome (trisomy 13)	38	2.0	34	1.9
Edwards syndrome (trisomy 18)	84	4.4	82	4.6
Cri du Chat syndrome	3	0.2	5	0.3

* Rate = (number of birth defects)/(number of live births)x10,000.
+ Birth weight >2,499 grams.

TABLE 3. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY GENDER, HAWAI'I, 1986-2005

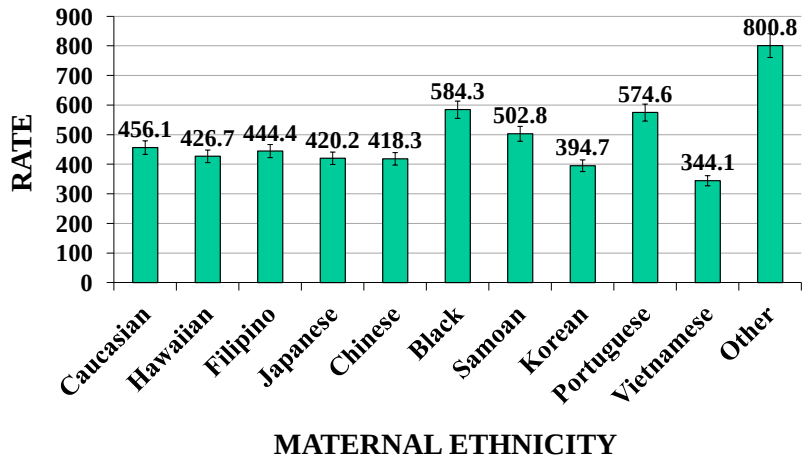
Diagnosis	Male		Female	
	N	Rate*	N	Rate
Wolff-Hirschorn syndrome	3	0.2	5	0.3
Autosomal anomalies in normal infant	63	3.3	77	4.3
Triploidy	21	1.1	19	1.1
Turner syndrome (45,X)	4	0.2	142	7.9
Klinefelter syndrome (47,XXY)	54	2.8	0	0.0
Absent spleen/Asplenia	3	0.2	10	0.6
Accessory spleen	28	1.5	14	0.8
Situs inversus	23	1.2	20	1.1
Conjoined twins	5	0.3	0	0.0
Tuberous sclerosis	7	0.4	8	0.4
Amniotic band syndrome	42	2.2	34	1.9
DiGeorge syndrome	15	0.8	10	0.6
Pierre-Robin sequence	20	1.0	16	0.9
Werdnig-Hoffman syndrome	9	0.5	6	0.3
Wolff-Parkinson-White syndrome	11	0.6	8	0.4
NEOPLASMS	158	8.3	202	11.2
TOTAL BIRTH DEFECT CASES	10,030	525.5	6,932	386.0
FETAL ALCOHOL SYNDROME	50	2.6	38	2.1
BIRTH DEFECTS WITH:				
CONGENITAL INFECTION	145	7.6	121	6.7
MATERNAL ALCOHOL USE	816	42.8	685	38.1
MATERNAL TOBACCO USE	2,726	142.8	2,223	123.8
MATERNAL ILLICIT DRUG USE	1,602	83.9	1,532	85.3
Marijuana	500	26.2	492	27.4
Cocaine	302	15.8	275	15.3
Methamphetamine/Amphetamine	1,068	56.0	1,037	57.8

* Rate = (number of birth defects)/(number of live births)x10,000.
+ Birth weight >2,499 grams.

Birth Defect Rates by Maternal Ethnicity[8]

There is substantial variation in birth defect rates among different maternal ethnic groups (Figure 4). The highest rates were in Black (584.3 per 10,000 live births), Portuguese (574.6), Samoan (502.8), and "Other"[9] (800.8) women, and the lowest rate was in Vietnamese women (344.1).

FIGURE 4. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY MATERNAL ETHNICITY, HAWAI'I, 1986-2005



There were also differences among broad categories and specific birth defects by maternal ethnicity (Table 4).

[8] A single ethnicity is assigned to each mother according to guidelines of the Hawai'i State Department of Health Office of Health Status Monitoring:

1. If the woman is noted to be of a single ethnicity, that ethnicity is listed.
2. If Hawaiian is one of the multiple ethnicities listed, Part-Hawaiian is coded.
3. If a non-Caucasian ethnicity is listed with Caucasian ethnicity, the non-Caucasian ethnicity is coded.
4. If there is more than one non-Caucasian ethnicity listed, the first one is coded.
5. If there is more than one Caucasian ethnicity listed, the first one is coded.

[9] The "Other" category includes American Indian, Asian Indian, Puerto Rican, Mexican, Micronesian, Tongan, and Other Pacific Islander.

TABLE 4. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY MATERNAL ETHNICITY, HAWAI‘I, 1986-2005

Diagnosis	Caucasian N=89,186		Hawaiian N=94,325		Filipino N=68,898		Japanese N=45,620		Chinese N=13,698		Black N=11,787	
	N	Rate*	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
BRAIN & NERVOUS SYSTEM	346	38.8	411	43.6	271	39.3	153	33.5	46	33.6	46	39.1
Neural tube defects	96	10.8	83	8.8	55	8.0	43	9.4	11	8.0	7	6.0
Anencephaly	32	3.6	34	3.6	17	2.5	17	3.7	5	3.7	1	0.9
Spina bifida	47	5.3	37	3.9	23	3.3	15	3.3	5	3.7	3	2.6
Encephalocele	17	1.9	15	1.6	16	2.3	11	2.4	2	1.5	4	3.4
Microcephaly	73	8.2	110	11.7	85	12.3	28	6.1	8	5.8	13	11.1
Holoprosencephaly	4	0.4	10	1.1	11	1.6	5	1.1	3	2.2	1	0.9
Hydrocephaly (w/o spina bifida)	93	10.4	111	11.8	69	10.0	36	7.9	10	7.3	15	12.8
EYE, EAR, FACE, & NECK	420	47.1	478	50.7	371	53.8	196	43.0	68	49.6	49	41.7
EYE	177	19.8	180	19.1	133	19.3	72	15.8	24	17.5	11	9.4
Anophthalmia	1	0.1	5	0.5	2	0.3	1	0.2	0	0.0	0	0.0
Microphthalmia	32	3.6	27	2.9	11	1.6	12	2.6	6	4.4	1	0.9
Cataract	14	1.6	12	1.3	4	0.6	4	0.9	1	0.7	3	2.6
EAR	295	33.1	335	35.5	277	40.2	138	30.2	49	35.8	37	31.5
Absent auditory canal/ear	4	0.4	14	1.5	11	1.6	4	0.9	1	0.7	0	0.0
Microtia	18	2.0	35	3.7	28	4.1	13	2.8	6	4.4	2	1.7
CARDIAC & CIRCULATORY	1,440	161.5	1,779	188.6	1,406	204.1	734	160.9	222	162.1	260	221.1
Common truncus	6	0.7	3	0.3	6	0.9	5	1.1	1	0.7	3	2.6
Transposition of the great vessels	24	2.7	38	4.0	18	2.6	11	2.4	3	2.2	0	0.0
Tetralogy of Fallot	19	2.1	47	5.0	28	4.1	23	5.0	3	2.2	6	5.1
Single ventricle	7	0.8	8	0.8	4	0.6	3	0.7	0	0.0	1	0.9
Ventricular septal defect	347	38.9	379	40.2	283	41.1	192	42.1	55	40.2	51	43.4
Atrial septal defect	184	20.6	257	27.2	184	26.7	103	22.6	21	15.3	31	26.4
Endocardial cushion defect	25	2.8	26	2.8	11	1.6	8	1.8	6	4.4	2	1.7
Pulmonary valve atresia/stenosis	59	6.6	98	10.4	65	9.4	55	12.1	10	7.3	10	8.5
Tricuspid valve atresia/stenosis	11	1.2	25	2.7	6	0.9	5	1.1	3	2.2	1	0.9
Ebstein’s anomaly	6	0.7	2	0.2	2	0.3	4	0.9	0	0.0	0	0.0
Aortic valve atresia/stenosis	13	1.5	7	0.7	7	1.0	10	2.2	3	2.2	2	1.7
Mitral valve atresia/stenosis	7	0.8	12	1.3	5	0.7	7	1.5	1	0.7	0	0.0
Hypoplastic left heart syndrome	15	1.7	13	1.4	8	1.2	6	1.3	2	1.5	1	0.9
Dextrocardia	17	1.9	15	1.6	2	0.3	5	1.1	3	2.2	3	2.6
Patent ductus arteriosus +	545	61.1	750	79.5	627	91.0	282	61.8	105	76.7	98	83.4
Coarctation of the aorta	37	4.1	18	1.9	22	3.2	9	2.0	1	0.7	3	2.6

* Rate = (number of birth defects)/(number of live births)x10,000.
+ Birth weight >2,499 grams.

TABLE 4. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY MATERNAL ETHNICITY, HAWAI‘I, 1986-2005

Diagnosis	Caucasian N=89,186		Hawaiian N=94,325		Filipino N=68,898		Japanese N=45,620		Chinese N=13,698		Black N=11,787	
	N	Rate*	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Aortic stenosis	1	0.1	0	0.0	1	0.1	0	0.0	0	0.0	0	0.0
Pulmonary artery atresia/stenosis	127	14.2	192	20.4	159	23.1	63	13.8	21	15.3	44	37.4
Total anomalous pulmonary venous return	5	0.6	6	0.6	7	1.0	8	1.8	3	2.2	2	1.7
Partial anomalous pulmonary venous return	1	0.1	4	0.4	3	0.4	3	0.7	0	0.0	2	1.7
Persistent fetal circulation	50	5.6	49	5.2	47	6.8	19	4.2	9	6.6	7	6.0
RESPIRATORY	238	26.7	223	23.6	181	26.3	89	19.5	31	22.6	43	36.6
Choanal atresia/stenosis	14	1.6	9	1.0	13	1.9	4	0.9	3	2.2	3	2.6
Lung cysts	3	0.3	14	1.5	8	1.2	2	0.4	1	0.7	0	0.0
Hypoplastic lung	56	6.3	46	4.9	38	5.5	28	6.1	5	3.7	10	8.5
OROFACIAL & GASTROINTESTINAL	564	63.2	502	53.2	375	54.4	278	60.9	84	61.3	73	62.1
Oral clefts	136	15.2	184	19.5	135	19.6	113	24.8	36	26.3	17	14.5
Cleft palate (isolated)	52	5.8	85	9.0	41	6.0	41	9.0	15	11.0	3	2.6
Cleft lip (+/-cleft palate)	84	9.4	99	10.5	94	13.6	72	15.8	22	16.1	14	11.9
TE fistula/Esophageal atresia	25	2.8	17	1.8	12	1.7	7	1.5	2	1.5	3	2.6
Pyloric stenosis	139	15.6	46	4.9	15	2.2	34	7.5	5	3.7	13	11.1
Atresia/stenosis small intestine	24	2.7	27	2.9	14	2.0	23	5.0	3	2.2	2	1.7
Anal atresia/stenosis	40	4.5	44	4.7	36	5.2	22	4.8	10	7.3	7	6.0
Hirschsprung’s disease	17	1.9	13	1.4	19	2.8	11	2.4	7	5.1	8	6.8
Malrotation	19	2.1	27	2.9	18	2.6	12	2.6	4	2.9	4	3.4
Biliary atresia	6	0.7	10	1.1	8	1.2	5	1.1	1	0.7	0	0.0
GENITAL & URINARY	995	111.6	1,002	106.2	811	117.7	362	79.4	110	80.3	185	157.4
Ovarian cyst	7	0.8	3	0.3	3	0.4	4	0.9	0	0.0	1	0.9
Undescended testicle +	275	30.8	330	35.0	250	36.3	92	20.2	20	14.6	58	49.3
Hypospadias/Epispadias/Chordee	326	36.6	283	30.0	271	39.3	109	23.9	32	23.4	51	43.4
Ambiguous genitalia	4	0.4	5	0.5	5	0.7	1	0.2	1	0.7	2	1.7
Absent/hypoplastic kidney	46	5.2	37	3.9	40	5.8	13	2.8	8	5.8	6	5.1
Cystic kidney	41	4.6	35	3.7	24	3.5	18	3.9	4	2.9	10	8.5
Hydronephrosis	138	15.5	109	11.6	84	12.2	49	10.7	19	13.9	23	19.6
Exstrophy bladder	2	0.2	1	0.1	2	0.3	1	0.2	0	0.0	0	0.0
Posterior urethral valves	8	0.9	5	0.5	1	0.1	2	0.4	0	0.0	3	2.6
LIMB & MUSCULOSKELETAL	1,323	148.3	1,141	121.0	784	113.8	532	116.6	166	121.2	222	188.8
Scoliosis	20	2.2	23	2.4	16	2.3	13	2.8	3	2.2	1	0.9
Hip dislocation	122	13.7	66	7.0	63	9.1	71	15.6	18	13.1	1	0.9

* Rate = (number of birth defects)/(number of live births)x10,000.
+ Birth weight >2,499 grams.

TABLE 4. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY MATERNAL ETHNICITY, HAWAI‘I, 1986-2005

Diagnosis	Caucasian N=89,186		Hawaiian N=94,325		Filipino N=68,898		Japanese N=45,620		Chinese N=13,698		Black N=11,787	
	N	Rate*	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Clubfoot	337	37.8	349	37.0	163	23.7	93	20.4	31	22.6	42	35.7
Polydactyly	111	12.4	140	14.8	75	10.9	60	13.2	28	20.4	105	89.3
Syndactyly	101	11.3	77	8.2	36	5.2	26	5.7	8	5.8	6	5.1
Limb reduction deformity - upper	26	2.9	38	4.0	27	3.9	10	2.2	4	2.9	5	4.3
Limb reduction deformity - lower	9	1.0	16	1.7	11	1.6	7	1.5	1	0.7	2	1.7
Craniosynostosis	71	8.0	33	3.5	26	3.8	25	5.5	6	4.4	4	3.4
Crouzon’s disease	0	0.0	1	0.1	1	0.1	0	0.0	0	0.0	0	0.0
Apert syndrome	3	0.3	2	0.2	3	0.4	1	0.2	1	0.7	0	0.0
Goldenhar syndrome	3	0.3	6	0.6	3	0.4	3	0.7	1	0.7	1	0.9
Spina bifida occulta	6	0.7	2	0.2	2	0.3	2	0.4	0	0.0	0	0.0
Dwarfism	26	2.9	20	2.1	21	3.0	19	4.2	1	0.7	0	0.0
Osteogenesis imperfecta	5	0.6	4	0.4	2	0.3	1	0.2	0	0.0	0	0.0
Diaphragmatic hernia	28	3.1	19	2.0	16	2.3	10	2.2	1	0.7	1	0.9
Abdominal wall defects	89	10.0	73	7.7	59	8.6	29	6.4	7	5.1	13	11.1
Omphalocele	27	3.0	17	1.8	20	2.9	10	2.2	5	3.7	3	2.6
Gastroschisis	30	3.4	38	4.0	22	3.2	12	2.6	2	1.5	7	6.0
Prune-belly syndrome	4	0.4	1	0.1	3	0.4	0	0.0	0	0.0	0	0.0
Poland syndrome	2	0.2	1	0.1	2	0.3	0	0.0	0	0.0	0	0.0
SKIN & INTEGUMENT	309	34.6	604	64.0	490	71.1	279	61.2	88	64.2	125	106.3
CHROMOSOME & SYNDROME	526	59.0	331	35.1	340	49.3	345	75.6	105	76.7	50	42.5
Down syndrome (trisomy 21)	148	16.6	89	9.4	100	14.5	85	18.6	36	26.3	6	5.1
Patau syndrome (trisomy 13)	22	2.5	9	1.0	8	1.2	11	2.4	7	5.1	2	1.7
Edwards syndrome (trisomy 18)	47	5.3	22	2.3	25	3.6	29	6.4	9	6.6	3	2.6
Cri du Chat syndrome	3	0.3	0	0.0	1	0.1	0	0.0	1	0.7	0	0.0
Wolff-Hirschorn syndrome	1	0.1	0	0.0	2	0.3	3	0.7	0	0.0	0	0.0
Autosomal anomalies in normal infant	35	3.9	19	2.0	32	4.6	30	6.6	11	8.0	3	2.6
Triploidy	11	1.2	5	0.5	3	0.4	7	1.5	3	2.2	1	0.9
Turner syndrome (45,X)	34	3.8	21	2.2	21	3.0	33	7.2	7	5.1	7	6.0
Klinefelter syndrome (47,XXY)	10	1.1	6	0.6	11	1.6	17	3.7	2	1.5	1	0.9
Absent spleen/Asplenia	2	0.2	6	0.6	1	0.1	1	0.2	1	0.7	0	0.0
Accessory spleen	9	1.0	11	1.2	9	1.3	6	1.3	1	0.7	0	0.0
Situs inversus	7	0.8	14	1.5	7	1.0	5	1.1	1	0.7	0	0.0
Conjoined twins	2	0.2	2	0.2	3	0.4	0	0.0	0	0.0	0	0.0

* Rate = (number of birth defects)/(number of live births)x10,000.
+ Birth weight >2,499 grams.

TABLE 4. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY MATERNAL ETHNICITY, HAWAI'I, 1986-2005

Diagnosis	Caucasian N=89,186		Hawaiian N=94,325		Filipino N=68,898		Japanese N=45,620		Chinese N=13,698		Black N=11,787	
	N	Rate*	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Tuberous sclerosis	4	0.4	4	0.4	3	0.4	1	0.2	1	0.7	1	0.9
Amniotic band syndrome	22	2.5	25	2.7	11	1.6	7	1.5	1	0.7	4	3.4
DiGeorge syndrome	5	0.6	9	1.0	2	0.3	4	0.9	0	0.0	1	0.9
Pierre-Robin sequence	7	0.8	11	1.2	6	0.9	6	1.3	2	1.5	1	0.9
Werdnig-Hoffman syndrome	3	0.3	4	0.4	0	0.0	1	0.2	0	0.0	1	0.9
Wolff-Parkinson-White syndrome	8	0.9	5	0.5	3	0.4	1	0.2	1	0.7	0	0.0
NEOPLASMS	82	9.2	78	8.3	67	9.7	55	12.1	14	10.2	13	11.1
TOTAL BIRTH DEFECT CASES	4,068	456.1	4,025	426.7	3,062	444.4	1,917	420.2	573	418.3	687	584.3
FETAL ALCOHOL SYNDROME	26	2.9	42	4.5	6	0.9	4	0.9	1	0.7	1	0.9
BIRTH DEFECTS CASES WITH:												
CONGENITAL INFECTION	53	5.9	59	6.3	60	8.7	26	5.7	10	7.3	13	11.1
MATERNAL ALCOHOL USE	431	48.3	579	61.4	125	18.1	119	26.1	18	13.1	53	45.1
MATERNAL TOBACCO USE	911	102.1	2,236	237.1	521	75.6	378	82.9	44	32.1	95	80.8
MATERNAL ILLICIT DRUG USE	404	45.3	1,729	183.3	348	50.5	187	41.0	27	19.7	48	40.8
Marijuana	162	18.2	552	58.5	79	11.5	41	9.0	9	6.6	20	17.0
Cocaine	127	14.2	282	29.9	28	4.1	34	7.5	7	5.1	16	13.6
Methamphetamine/Amphetamine	173	19.4	1,231	130.5	275	39.9	141	30.9	15	11.0	21	17.9

* Rate = (number of birth defects)/(number of live births)x10,000.
+ Birth weight >2,499 grams.

TABLE 4. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY MATERNAL ETHNICITY, HAWAI‘I, 1986-2005

Diagnosis	Samoan N=10,620		Korean N=7,474		Portuguese N=3,202		Vietnamese N=2,906		Other N=7,466	
	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
BRAIN & NERVOUS SYSTEM	54	50.8	25	33.4	22	68.7	8	27.5	113	66.0
Neural tube defects	14	13.2	4	5.4	4	12.5	2	6.9	22	12.8
Anencephaly	3	2.8	4	5.4	1	3.1	1	3.4	6	3.5
Spina bifida	8	7.5	0	0.0	3	9.4	1	3.4	14	8.2
Encephalocele	3	2.8	0	0.0	1	3.1	0	0.0	4	2.3
Microcephaly	7	6.6	8	10.7	5	15.6	0	0.0	26	15.2
Holoprosencephaly	0	0.0	3	4.0	0	0.0	1	3.4	6	3.5
Hydrocephaly (w/o spina bifida)	14	13.2	6	8.0	4	12.5	1	3.4	31	18.1
EYE, EAR, FACE, & NECK	58	54.6	40	53.5	21	65.6	8	27.5	162	94.6
EYE	17	16.0	15	20.1	9	28.1	3	10.3	65	37.9
Anophthalmia	1	0.9	0	0.0	0	0.0	0	0.0	3	1.8
Microphthalmia	3	2.8	2	2.7	0	0.0	0	0.0	12	7.0
Cataract	0	0.0	1	1.3	0	0.0	0	0.0	6	3.5
EAR	43	40.5	27	36.1	15	46.8	4	13.8	102	59.5
Absent auditory canal/ear	3	2.8	2	2.7	0	0.0	0	0.0	5	2.9
Microtia	9	8.5	3	4.0	2	6.2	0	0.0	13	7.6
CARDIAC & CIRCULATORY	259	243.9	106	141.8	69	215.5	50	172.1	660	385.2
Common truncus	1	0.9	0	0.0	0	0.0	1	3.4	1	0.6
Transposition of the great vessels	1	0.9	2	2.7	1	3.1	1	3.4	9	5.3
Tetralogy of Fallot	1	0.9	4	5.4	1	3.1	1	3.4	7	4.1
Single ventricle	0	0.0	2	2.7	0	0.0	0	0.0	3	1.8
Ventricular septal defect	47	44.3	30	40.1	11	34.4	12	41.3	114	66.5
Atrial septal defect	25	23.5	23	30.8	8	25.0	5	17.2	92	53.7
Endocardial cushion defect	1	0.9	0	0.0	0	0.0	0	0.0	7	4.1
Pulmonary valve atresia/stenosis	7	6.6	6	8.0	1	3.1	3	10.3	32	18.7
Tricuspid valve atresia/stenosis	1	0.9	0	0.0	0	0.0	1	3.4	5	2.9
Ebstein’s anomaly	1	0.9	0	0.0	0	0.0	0	0.0	3	1.8
Aortic valve atresia/stenosis	3	2.8	2	2.7	2	6.2	0	0.0	3	1.8
Mitral valve atresia/stenosis	2	1.9	0	0.0	1	3.1	0	0.0	3	1.8
Hypoplastic left heart syndrome	2	1.9	2	2.7	1	3.1	0	0.0	5	2.9
Dextrocardia	3	2.8	2	2.7	0	0.0	0	0.0	3	1.8
Patent ductus arteriosus +	140	131.8	46	61.5	30	93.7	24	82.6	340	198.4
Coarctation of the aorta	1	0.9	2	2.7	1	3.1	1	3.4	7	4.1

* Rate = (number of birth defects)/(number of live births)x10,000.
+ Birth weight >2,499 grams.

TABLE 4. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY MATERNAL ETHNICITY, HAWAI‘I, 1986-2005

Diagnosis	Samoan N=10,620		Korean N=7,474		Portuguese N=3,202		Vietnamese N=2,906		Other N=7,466	
	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Aortic stenosis	1	0.9	0	0.0	0	0.0	0	0.0	0	0.0
Pulmonary artery atresia/stenosis	31	29.2	5	6.7	3	9.4	9	31.0	70	40.9
Total anomalous pulmonary venous return	1	0.9	1	1.3	0	0.0	0	0.0	4	2.3
Partial anomalous pulmonary venous return	1	0.9	0	0.0	0	0.0	0	0.0	3	1.8
Persistent fetal circulation	15	14.1	1	1.3	1	3.1	1	3.4	14	8.2
RESPIRATORY	26	24.5	23	30.8	17	53.1	3	10.3	55	32.1
Choanal atresia/stenosis	0	0.0	2	2.7	1	3.1	0	0.0	1	0.6
Lung cysts	2	1.9	0	0.0	0	0.0	0	0.0	1	0.6
Hypoplastic lung	6	5.6	4	5.4	2	6.2	0	0.0	16	9.3
OROFACIAL & GASTROINTESTINAL	71	66.9	49	65.6	27	84.3	15	51.6	154	89.9
Oral clefts	22	20.7	19	25.4	10	31.2	6	20.6	45	26.3
Cleft palate (isolated)	6	5.6	6	8.0	3	9.4	1	3.4	16	9.3
Cleft lip (+/-cleft palate)	16	15.1	13	17.4	7	21.9	5	17.2	29	16.9
TE fistula/Esophageal atresia	1	0.9	2	2.7	0	0.0	0	0.0	8	4.7
Pyloric stenosis	0	0.0	7	9.4	5	15.6	1	3.4	16	9.3
Atresia/stenosis small intestine	2	1.9	2	2.7	0	0.0	1	3.4	7	4.1
Anal atresia/stenosis	6	5.6	2	2.7	1	3.1	1	3.4	11	6.4
Hirschsprung’s disease	5	4.7	2	2.7	1	3.1	0	0.0	9	5.3
Malrotation	6	5.6	0	0.0	1	3.1	0	0.0	9	5.3
Biliary atresia	2	1.9	0	0.0	2	6.2	0	0.0	6	3.5
GENITAL & URINARY	130	122.4	71	95.0	39	121.8	18	61.9	316	184.4
Ovarian cyst	0	0.0	0	0.0	0	0.0	0	0.0	1	0.6
Undescended testicle +	31	29.2	18	24.1	16	50.0	7	24.1	146	85.2
Hypospadias/Epispadias/Chordee	38	35.8	28	37.5	9	28.1	5	17.2	94	54.9
Ambiguous genitalia	0	0.0	0	0.0	0	0.0	0	0.0	2	1.2
Absent/hypoplastic kidney	5	4.7	1	1.3	0	0.0	0	0.0	10	5.8
Cystic kidney	10	9.4	2	2.7	1	3.1	0	0.0	14	8.2
Hydronephrosis	18	16.9	7	9.4	7	21.9	1	3.4	48	28.0
Exstrophy bladder	0	0.0	1	1.3	0	0.0	1	3.4	1	0.6
Posterior urethral valves	1	0.9	1	1.3	0	0.0	0	0.0	1	0.6
LIMB & MUSCULOSKELETAL	151	142.2	84	112.4	49	153.0	32	110.1	389	227.0
Scoliosis	2	1.9	1	1.3	1	3.1	0	0.0	8	4.7
Hip dislocation	5	4.7	7	9.4	1	3.1	1	3.4	18	10.5

* Rate = (number of birth defects)/(number of live births)x10,000.
+ Birth weight >2,499 grams.

TABLE 4. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY MATERNAL ETHNICITY, HAWAI‘I, 1986-2005

Diagnosis	Samoan N=10,620		Korean N=7,474		Portuguese N=3,202		Vietnamese N=2,906		Other N=7,466	
	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Clubfoot	46	43.3	13	17.4	17	53.1	3	10.3	102	59.5
Polydactyly	26	24.5	12	16.1	4	12.5	7	24.1	68	39.7
Syndactyly	8	7.5	1	1.3	1	3.1	3	10.3	28	16.3
Limb reduction deformity - upper	5	4.7	3	4.0	2	6.2	1	3.4	8	4.7
Limb reduction deformity - lower	3	2.8	2	2.7	1	3.1	1	3.4	0	0.0
Craniosynostosis	0	0.0	2	2.7	2	6.2	0	0.0	11	6.4
Crouzon’s disease	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Apert syndrome	0	0.0	0	0.0	0	0.0	0	0.0	1	0.6
Goldenhar syndrome	1	0.9	0	0.0	0	0.0	0	0.0	3	1.8
Spina bifida occulta	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Dwarfism	1	0.9	5	6.7	1	3.1	0	0.0	7	4.1
Osteogenesis imperfecta	1	0.9	0	0.0	0	0.0	1	3.4	1	0.6
Diaphragmatic hernia	4	3.8	1	1.3	0	0.0	2	6.9	3	1.8
Abdominal wall defects	5	4.7	9	12.0	4	12.5	2	6.9	23	13.4
Omphalocele	1	0.9	6	8.0	0	0.0	1	3.4	6	3.5
Gastroschisis	4	3.8	3	4.0	1	3.1	1	3.4	10	5.8
Prune-belly syndrome	0	0.0	0	0.0	0	0.0	0	0.0	2	1.2
Poland syndrome	0	0.0	0	0.0	0	0.0	0	0.0	1	0.6
SKIN & INTEGUMENT	92	86.6	51	68.2	22	68.7	20	68.8	201	117.3
CHROMOSOME & SYNDROME	42	39.5	47	62.9	19	59.3	7	24.1	130	75.9
Down syndrome (trisomy 21)	8	7.5	11	14.7	6	18.7	2	6.9	35	20.4
Patau syndrome (trisomy 13)	1	0.9	4	5.4	1	3.1	0	0.0	5	2.9
Edwards syndrome (trisomy 18)	2	1.9	3	4.0	1	3.1	1	3.4	8	4.7
Cri du Chat syndrome	2	1.9	0	0.0	0	0.0	0	0.0	0	0.0
Wolff-Hirschorn syndrome	0	0.0	1	1.3	0	0.0	0	0.0	0	0.0
Autosomal anomalies in normal infant	1	0.9	5	6.7	0	0.0	0	0.0	4	2.3
Triploidy	0	0.0	0	0.0	1	3.1	0	0.0	0	0.0
Turner syndrome (45,X)	1	0.9	2	2.7	2	6.2	1	3.4	8	4.7
Klinefelter syndrome (47,XXY)	0	0.0	2	2.7	0	0.0	0	0.0	5	2.9
Absent spleen/Asplenia	0	0.0	1	1.3	0	0.0	0	0.0	1	0.6
Accessory spleen	4	3.8	0	0.0	0	0.0	0	0.0	2	1.2
Situs inversus	1	0.9	3	4.0	0	0.0	0	0.0	3	1.8
Conjoined twins	2	1.9	2	2.7	2	6.2	0	0.0	0	0.0

* Rate = (number of birth defects)/(number of live births)x10,000.
+ Birth weight >2,499 grams.

TABLE 4. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY MATERNAL ETHNICITY, HAWAI‘I, 1986-2005

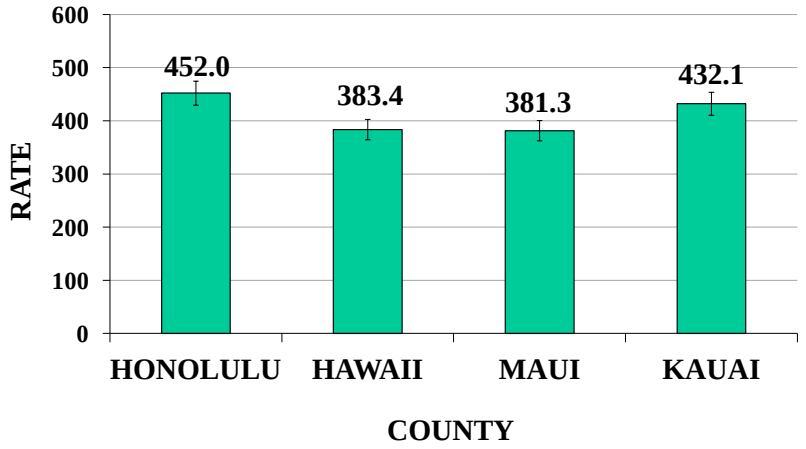
Diagnosis	Samoan N=10,620		Korean N=7,474		Portuguese N=3,202		Vietnamese N=2,906		Other N=7,466	
	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Tuberous sclerosis	0	0.0	1	1.3	0	0.0	0	0.0	0	0.0
Amniotic band syndrome	0	0.0	2	2.7	1	3.1	1	3.4	4	2.3
DiGeorge syndrome	0	0.0	3	4.0	0	0.0	0	0.0	1	0.6
Pierre-Robin sequence	0	0.0	0	0.0	1	3.1	0	0.0	0	0.0
Werdnig-Hoffman syndrome	3	2.8	0	0.0	0	0.0	0	0.0	3	1.8
Wolff-Parkinson-White syndrome	1	0.9	0	0.0	0	0.0	0	0.0	1	0.6
NEOPLASMS	10	9.4	9	12.0	6	18.7	0	0.0	23	13.4
TOTAL BIRTH DEFECT CASES	534	502.8	295	394.7	184	574.6	100	344.1	1,372	800.8
FETAL ALCOHOL SYNDROME	0	0.0	1	1.3	0	0.0	2	6.9	7	4.1
BIRTH DEFECTS CASES WITH:										
CONGENITAL INFECTION	12	11.3	1	1.3	2	6.2	1	3.4	28	16.3
MATERNAL ALCOHOL USE	34	32.0	19	25.4	20	62.5	3	10.3	12	7.0
MATERNAL TOBACCO USE	197	185.5	89	119.1	76	237.4	5	17.2	35	20.4
MATERNAL ILLICIT DRUG USE	81	76.3	48	64.2	45	140.5	4	13.8	219	127.8
Marijuana	29	27.3	11	14.7	18	56.2	1	3.4	77	44.9
Cocaine	10	9.4	9	12.0	17	53.1	3	10.3	53	30.9
Methamphetamine/Amphetamine	53	49.9	36	48.2	25	78.1	1	3.4	131	76.5

* Rate = (number of birth defects)/(number of live births)x10,000.
+ Birth weight >2,499 grams.

Birth Defect Rates by County[10]

The birth defect rates for Hawai‘i County (383.4 per 10,000 live births) and Maui County (381.3) were similar and significantly lower compared to the rates for the City and County of Honolulu (452.0) and Kauai County (432.1) (Figure 5).

FIGURE 5. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY COUNTY, HAWAI‘I, 1986-2005



There were also differences among broad categories and specific birth defects by county (Table 5).

[10] Based on maternal residence.

TABLE 5. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY COUNTY, HAWAI‘I, 1986-2005

Diagnosis	Honolulu N=289,646		Hawaii N=42,797		Maui N=37,186		Kauai N=17,633	
	N	Rate*	N	Rate	N	Rate	N	Rate
BRAIN & NERVOUS SYSTEM	1,108	38.3	205	47.9	124	33.3	67	38.0
Neural tube defects	263	9.1	37	8.6	35	9.4	12	6.8
Anencephaly	101	3.5	12	2.8	13	3.5	5	2.8
Spina bifida	115	4.0	14	3.3	19	5.1	5	2.8
Encephalocele	56	1.9	11	2.6	3	0.8	3	1.7
Microcephaly	263	9.1	55	12.9	25	6.7	22	12.5
Holoprosencephaly	31	1.1	7	1.6	7	1.9	2	1.1
Hydrocephaly (w/o spina bifida)	290	10.0	46	10.7	35	9.4	14	7.9
EYE, EAR, FACE, & NECK	1,359	46.9	232	54.2	189	50.8	78	44.2
EYE	481	16.6	111	25.9	70	18.8	31	17.6
Anophthalmia	8	0.3	3	0.7	3	0.8	0	0.0
Microphthalmia	71	2.5	16	3.7	12	3.2	6	3.4
Cataract	30	1.0	11	2.6	2	0.5	0	0.0
EAR	961	33.2	154	36.0	137	36.8	57	32.3
Absent auditory canal/ear	32	1.1	6	1.4	6	1.6	1	0.6
Microtia	101	3.5	11	2.6	11	3.0	6	3.4
CARDIAC & CIRCULATORY	5,421	187.2	685	160.1	542	145.8	332	188.3
Common truncus	22	0.8	1	0.2	1	0.3	2	1.1
Transposition of the great vessels	110	3.8	25	5.8	18	4.8	6	3.4
Tetralogy of Fallot	107	3.7	13	3.0	13	3.5	5	2.8
Single ventricle	19	0.7	3	0.7	5	1.3	1	0.6
Ventricular septal defect	1,353	46.7	170	39.7	137	36.8	77	43.7
Atrial septal defect	2,022	69.8	285	66.6	257	69.1	135	76.6
Endocardial cushion defect	61	2.1	13	3.0	7	1.9	3	1.7
Pulmonary valve atresia/stenosis	258	8.9	41	9.6	30	8.1	12	6.8
Tricuspid valve atresia/stenosis	48	1.7	7	1.6	1	0.3	1	0.6
Ebstein’s anomaly	19	0.7	0	0.0	0	0.0	0	0.0
Aortic valve atresia/stenosis	35	1.2	10	2.3	6	1.6	1	0.6
Mitral valve atresia/stenosis	24	0.8	7	1.6	6	1.6	1	0.6
Hypoplastic left heart syndrome	36	1.2	11	2.6	10	2.7	1	0.6
Dextrocardia	34	1.2	8	1.9	10	2.7	1	0.6
Patent ductus arteriosus +	2,415	83.4	225	52.6	240	64.5	124	70.3
Coarctation of the aorta	71	2.5	13	3.0	16	4.3	5	2.8
Aortic stenosis	1	0.0	0	0.0	2	0.5	0	0.0
Pulmonary artery atresia/stenosis	563	19.4	79	18.5	50	13.4	33	18.7
Total anomalous pulmonary venous return	26	0.9	4	0.9	4	1.1	3	1.7
Partial anomalous pulmonary venous return	11	0.4	1	0.2	3	0.8	1	0.6
Persistent fetal circulation	146	5.0	23	5.4	28	7.5	17	9.6
RESPIRATORY	672	23.2	116	27.1	92	24.7	41	23.3
Choanal atresia/stenosis	36	1.2	7	1.6	6	1.6	1	0.6
Lung cysts	27	0.9	2	0.5	1	0.3	1	0.6
Hypoplastic lung	154	5.3	22	5.1	15	4.0	10	5.7
OROFACIAL & GASTROINTESTINAL	1,638	56.6	253	59.1	204	54.9	96	54.4
Oral clefts	525	18.1	93	21.7	77	20.7	31	17.6

* Rate = (number of birth defects)/(number of live births)x10,000.
\+ Birth weight >2,499 grams.

TABLE 5. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY COUNTY, HAWAI‘I, 1986-2005

Diagnosis	Honolulu N=289,646		Hawaii N=42,797		Maui N=37,186		Kauai N=17,633	
	N	Rate*	N	Rate	N	Rate	N	Rate
Cleft palate (isolated)	180	6.2	38	8.9	34	9.1	19	10.8
Cleft lip (+/-cleft palate)	346	11.9	55	12.9	43	11.6	12	6.8
TE fistula/Esophageal atresia	53	1.8	14	3.3	10	2.7	1	0.6
Pyloric stenosis	226	7.8	29	6.8	20	5.4	10	5.7
Atresia/stenosis small intestine	74	2.6	16	3.7	9	2.4	4	2.3
Anal atresia/stenosis	139	4.8	16	3.7	18	4.8	8	4.5
Hirschsprung’s disease	76	2.6	10	2.3	3	0.8	3	1.7
Malrotation	80	2.8	8	1.9	6	1.6	3	1.7
Biliary atresia	25	0.9	5	1.2	3	0.8	5	2.8
GENITAL & URINARY	3,064	105.8	379	88.6	393	105.7	189	107.2
Ovarian cyst	13	0.4	2	0.5	3	0.8	1	0.6
Undescended testicle +	871	30.1	113	26.4	130	35.0	51	28.9
Hypospadias/Epispadias/Chordee	931	32.1	116	27.1	138	37.1	62	35.2
Ambiguous genitalia	18	0.6	0	0.0	1	0.3	1	0.6
Absent/hypoplastic kidney	125	4.3	18	4.2	16	4.3	7	4.0
Cystic kidney	117	4.0	24	5.6	10	2.7	6	3.4
Hydronephrosis	399	13.8	43	10.0	42	11.3	19	10.8
Exstrophy bladder	5	0.2	0	0.0	4	1.1	0	0.0
Posterior urethral valves	18	0.6	2	0.5	2	0.5	0	0.0
LIMB & MUSCULOSKELETAL	3,727	128.7	500	116.8	410	110.3	221	125.3
Scoliosis	65	2.2	10	2.3	9	2.4	4	2.3
Hip dislocation	289	10.0	29	6.8	33	8.9	21	11.9
Clubfoot	868	30.0	141	32.9	120	32.3	58	32.9
Polydactyly	503	17.4	47	11.0	62	16.7	25	14.2
Syndactyly	219	7.6	38	8.9	23	6.2	14	7.9
Limb reduction deformity - upper	97	3.3	15	3.5	10	2.7	4	2.3
Limb reduction deformity - lower	43	1.5	3	0.7	3	0.8	5	2.8
Craniosynostosis	137	4.7	20	4.7	13	3.5	8	4.5
Crouzon’s disease	2	0.1	0	0.0	0	0.0	0	0.0
Apert syndrome	9	0.3	2	0.5	0	0.0	0	0.0
Goldenhar syndrome	13	0.4	3	0.7	2	0.5	1	0.6
Spina bifida occulta	7	0.2	3	0.7	1	0.3	2	1.1
Dwarfism	77	2.7	13	3.0	6	1.6	2	1.1
Osteogenesis imperfecta	9	0.3	2	0.5	2	0.5	2	1.1
Diaphragmatic hernia	67	2.3	6	1.4	8	2.2	4	2.3
Abdominal wall defects	235	8.1	33	7.7	31	8.3	15	8.5
Omphalocele	73	2.5	9	2.1	11	3.0	3	1.7
Gastroschisis	97	3.3	14	3.3	11	3.0	8	4.5
Prune-belly syndrome	8	0.3	0	0.0	1	0.3	1	0.6
Poland syndrome	5	0.2	1	0.2	0	0.0	0	0.0
SKIN & INTEGUMENT	1,929	66.6	180	42.1	106	28.5	60	34.0
CHROMOSOME & SYNDROME	1,576	54.4	207	48.4	159	42.8	83	47.1
Down syndrome (trisomy 21)	423	14.6	61	14.3	49	13.2	21	11.9
Patau syndrome (trisomy 13)	55	1.9	11	2.6	4	1.1	3	1.7

* Rate = (number of birth defects)/(number of live births)x10,000.
+ Birth weight >2,499 grams.

TABLE 5. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY COUNTY, HAWAI'I, 1986-2005

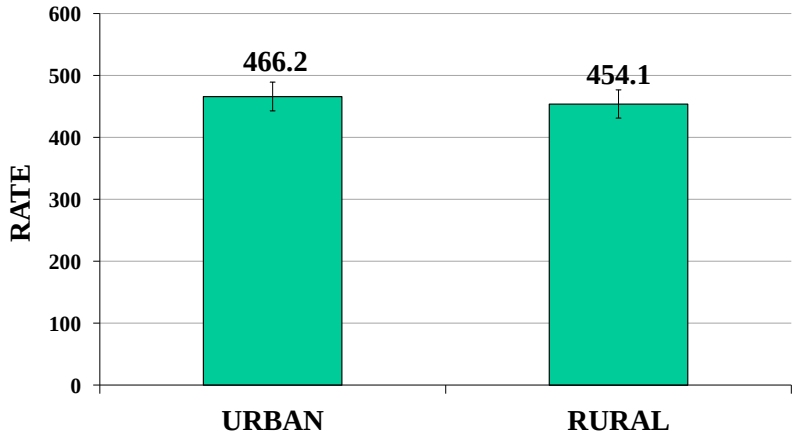
Diagnosis	Honolulu N=289,646		Hawaii N=42,797		Maui N=37,186		Kauai N=17,633	
	N	Rate*	N	Rate	N	Rate	N	Rate
Edwards syndrome (trisomy 18)	133	4.6	12	2.8	16	4.3	1	0.6
Cri du Chat syndrome	5	0.2	1	0.2	0	0.0	2	1.1
Wolff-Hirschorn syndrome	5	0.2	2	0.5	1	0.3	0	0.0
Autosomal anomalies in normal infant	119	4.1	7	1.6	5	1.3	8	4.5
Triploidy	35	1.2	1	0.2	2	0.5	2	1.1
Turner syndrome (45,X)	122	4.2	10	2.3	7	1.9	5	2.8
Klinefelter syndrome (47,XXY)	41	1.4	4	0.9	6	1.6	3	1.7
Absent spleen/Asplenia	9	0.3	2	0.5	1	0.3	0	0.0
Accessory spleen	28	1.0	6	1.4	4	1.1	2	1.1
Situs inversus	30	1.0	5	1.2	3	0.8	1	0.6
Conjoined twins	11	0.4	2	0.5	0	0.0	0	0.0
Tuberous sclerosis	13	0.4	1	0.2	1	0.3	0	0.0
Amniotic band syndrome	60	2.1	7	1.6	7	1.9	6	3.4
DiGeorge syndrome	17	0.6	6	1.4	2	0.5	0	0.0
Pierre-Robin sequence	23	0.8	6	1.4	5	1.3	2	1.1
Werdnig-Hoffman syndrome	11	0.4	2	0.5	3	0.8	0	0.0
Wolff-Parkinson-White syndrome	15	0.5	1	0.2	3	0.8	1	0.6
NEOPLASMS	297	10.3	31	7.2	27	7.3	12	6.8
TOTAL BIRTH DEFECT CASES	13,091	452.0	1,641	383.4	1,418	381.3	762	432.1
FETAL ALCOHOL SYNDROME	49	1.7	28	6.5	4	1.1	8	4.5
BIRTH DEFECTS CASES WITH:								
CONGENITAL INFECTION	201	6.9	24	5.6	17	4.6	21	11.9
MATERNAL ALCOHOL USE	1,144	39.5	175	40.9	111	29.8	78	44.2
MATERNAL TOBACCO USE	3,823	132.0	546	127.6	375	100.8	225	127.6
MATERNAL ILLICIT DRUG USE	2,508	86.6	339	79.2	211	56.7	117	66.4
Marijuana	754	26.0	139	32.5	74	19.9	43	24.4
Cocaine	396	13.7	110	25.7	51	13.7	32	18.1
Methamphetamine/Amphetamine	1,803	62.2	147	34.3	110	29.6	65	36.9

* Rate = (number of birth defects)/(number of live births)x10,000.
+ Birth weight >2,499 grams.

Birth Defect Rates by Urban/Rural Areas[11]

The birth defect rates for urban (466.2 per 10,000 live births) and rural (454.1) areas were similar (Figure 6).

FIGURE 6. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY URBAN/RURAL AREAS, HAWAI'I, 1986-2005



There were differences among broad categories and specific birth defects by urban and rural areas (Table 6).

[11] Based on maternal residence. Maternal residences in Metropolitan Honolulu (with zipcode starting with "968") were coded as "urban". Maternal residences in the rest of Hawai'i (with zipcode starting with "967") were coded as "rural".

TABLE 6. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY URBAN/RURAL AREAS, HAWAI‘I, 1986-2005

Diagnosis	Urban* N=109,522		Rural* N=259,842	
	N	Rate**	N	Rate
BRAIN & NERVOUS SYSTEM	432	39.4	1,073	41.3
Neural tube defects	106	9.7	240	9.2
Anencephaly	38	3.5	92	3.5
Spina bifida	50	4.6	103	4.0
Encephalocele	20	1.8	53	2.0
Microcephaly	100	9.1	265	10.2
Holoprosencephaly	13	1.2	34	1.3
Hydrocephaly (w/o spina bifida)	99	9.0	288	11.1
EYE, EAR, FACE, & NECK	549	50.1	1,309	50.4
EYE	194	17.7	499	19.2
Anophthalmia	5	0.5	9	0.3
Microphthalmia	34	3.1	71	2.7
Cataract	8	0.7	35	1.3
EAR	387	35.3	922	35.5
Absent auditory canal/ear	13	1.2	32	1.2
Microtia	42	3.8	87	3.3
CARDIAC & CIRCULATORY	2,124	193.9	4,855	186.8
Common truncus	10	0.9	16	0.6
Transposition of the great vessels	37	3.4	122	4.7
Tetralogy of Fallot	40	3.7	98	3.8
Single ventricle	6	0.5	22	0.8
Ventricular septal defect	563	51.4	1,174	45.2
Atrial septal defect	794	72.5	1,903	73.2
Endocardial cushion defect	23	2.1	61	2.3
Pulmonary valve atresia/stenosis	99	9.0	242	9.3
Tricuspid valve atresia/stenosis	17	1.6	40	1.5
Ebstein’s anomaly	6	0.5	13	0.5
Aortic valve atresia/stenosis	14	1.3	38	1.5
Mitral valve atresia/stenosis	7	0.6	31	1.2
Hypoplastic left heart syndrome	13	1.2	45	1.7
Dextrocardia	20	1.8	33	1.3
Patent ductus arteriosus +	934	85.3	2,069	79.6
Coarctation of the aorta	21	1.9	84	3.2
Aortic stenosis	1	0.1	2	0.1
Pulmonary artery atresia/stenosis	229	20.9	497	19.1
Total anomalous pulmonary venous return	13	1.2	23	0.9
Partial anomalous pulmonary venous return	4	0.4	12	0.5
Persistent fetal circulation	69	6.3	145	5.6
RESPIRATORY	267	24.4	654	25.2
Choanal atresia/stenosis	15	1.4	35	1.3
Lung cysts	12	1.1	19	0.7
Hypoplastic lung	65	5.9	136	5.2
OROFACIAL & GASTROINTESTINAL	634	57.9	1,556	59.9
Oral clefts	218	19.9	508	19.6

* Urban = Metropolitan Honolulu (any zipcode starting with 968). Rural = rest of Hawai‘i (any zipcode starting with 967).
** Rate = (number of birth defects)/(number of live births)x10,000.
\+ Birth weight >2,499 grams.

TABLE 6. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY URBAN/RURAL AREAS, HAWAI‘I, 1986-2005

Diagnosis	Urban* N=109,522		Rural* N=259,842	
	N	Rate**	N	Rate
Cleft palate (isolated)	74	6.8	197	7.6
Cleft lip (+/-cleft palate)	144	13.1	312	12.0
TE fistula/Esophageal atresia	20	1.8	58	2.2
Pyloric stenosis	73	6.7	212	8.2
Atresia/stenosis small intestine	28	2.6	75	2.9
Anal atresia/stenosis	55	5.0	126	4.8
Hirschsprung’s disease	35	3.2	57	2.2
Malrotation	29	2.6	68	2.6
Biliary atresia	12	1.1	26	1.0
GENITAL & URINARY	1,211	110.6	2,809	108.1
Ovarian cyst	5	0.5	14	0.5
Undescended testicle +	370	33.8	794	30.6
Hypospadias/Epispadias/Chordee	327	29.9	919	35.4
Ambiguous genitalia	6	0.5	14	0.5
Absent/hypoplastic kidney	53	4.8	113	4.3
Cystic kidney	54	4.9	103	4.0
Hydronephrosis	176	16.1	324	12.5
Exstrophy bladder	0	0.0	9	0.3
Posterior urethral valves	8	0.7	14	0.5
LIMB & MUSCULOSKELETAL	1,425	130.1	3,434	132.2
Scoliosis	26	2.4	62	2.4
Hip dislocation	104	9.5	268	10.3
Clubfoot	330	30.1	858	33.0
Polydactyly	185	16.9	452	17.4
Syndactyly	88	8.0	206	7.9
Limb reduction deformity - upper	34	3.1	93	3.6
Limb reduction deformity - lower	17	1.6	37	1.4
Craniosynostosis	46	4.2	132	5.1
Crouzon’s disease	1	0.1	1	0.0
Apert syndrome	3	0.3	8	0.3
Goldenhar syndrome	8	0.7	11	0.4
Spina bifida occulta	2	0.2	11	0.4
Dwarfism	27	2.5	71	2.7
Osteogenesis imperfecta	6	0.5	9	0.3
Diaphragmatic hernia	26	2.4	59	2.3
Abdominal wall defects	78	7.1	236	9.1
Omphalocele	28	2.6	68	2.6
Gastroschisis	30	2.7	100	3.8
Prune-belly syndrome	1	0.1	9	0.3
Poland syndrome	1	0.1	5	0.2
SKIN & INTEGUMENT	769	70.2	1,503	57.8
CHROMOSOME & SYNDROME	658	60.1	1,367	52.6
Down syndrome (trisomy 21)	192	17.5	362	13.9
Patau syndrome (trisomy 13)	25	2.3	48	1.8

* Urban = Metropolitan Honolulu (any zipcode starting with 968). Rural = rest of Hawai‘i (any zipcode starting with 967).
** Rate = (number of birth defects)/(number of live births)x10,000.
\+ Birth weight >2,499 grams.

TABLE 6. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY URBAN/RURAL AREAS, HAWAI‘I, 1986-2005

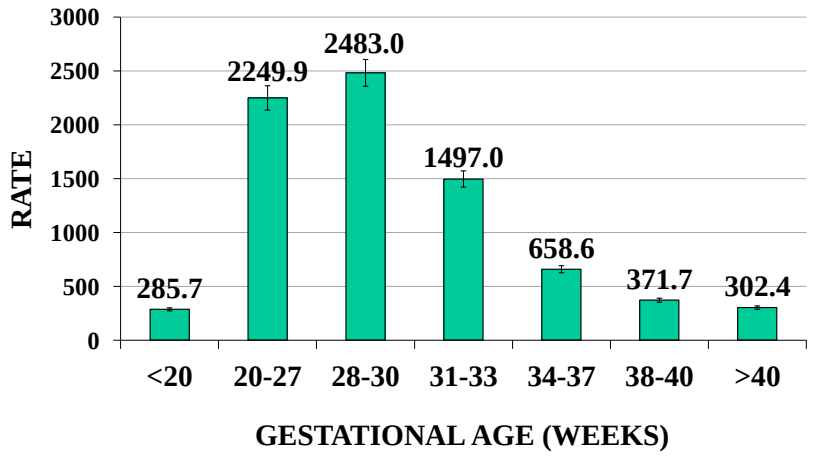
Diagnosis	Urban* N=109,522		Rural* N=259,842	
	N	Rate**	N	Rate
Edwards syndrome (trisomy 18)	66	6.0	96	3.7
Cri du Chat syndrome	2	0.2	5	0.2
Wolff-Hirschorn syndrome	3	0.3	5	0.2
Autosomal anomalies in normal infant	47	4.3	92	3.5
Triploidy	18	1.6	22	0.8
Turner syndrome (45,X)	51	4.7	93	3.6
Klinefelter syndrome (47,XXY)	16	1.5	38	1.5
Absent spleen/Asplenia	4	0.4	8	0.3
Accessory spleen	10	0.9	30	1.2
Situs inversus	15	1.4	24	0.9
Conjoined twins	4	0.4	9	0.3
Tuberous sclerosis	4	0.4	11	0.4
Amniotic band syndrome	24	2.2	57	2.2
DiGeorge syndrome	7	0.6	18	0.7
Pierre-Robin sequence	8	0.7	28	1.1
Werdnig-Hoffman syndrome	2	0.2	14	0.5
Wolff-Parkinson-White syndrome	4	0.4	16	0.6
NEOPLASMS	119	10.9	246	9.5
TOTAL BIRTH DEFECT CASES	5,106	466.2	11,799	454.1
FETAL ALCOHOL SYNDROME	26	2.4	63	2.4
BIRTH DEFECTS CASES WITH:				
CONGENITAL INFECTION	87	7.9	176	6.8
MATERNAL ALCOHOL USE	448	40.9	1,059	40.8
MATERNAL TOBACCO USE	1,238	113.0	3,724	143.3
MATERNAL ILLICIT DRUG USE	768	70.1	2,402	92.4
Marijuana	218	19.9	792	30.5
Cocaine	187	17.1	400	15.4
Methamphetamine/Amphetamine	504	46.0	1,617	62.2

* Urban = Metropolitan Honolulu (any zipcode starting with 968). Rural = rest of Hawai‘i (any zipcode starting with 967).
** Rate = (number of birth defects)/(number of live births)x10,000.
\+ Birth weight >2,499 grams.

Birth Defect Rates by Gestational Age

Birth defect rates for infants born early at 20-37 weeks gestational age were significantly higher than for infants born at ≥38 weeks (Figure 7). The rates were highest in the earliest gestational ages of 20-27 weeks (2,249.9 per 10,000 live births) and 28-30 weeks (2,483.0). The rate for gestational age <20 weeks was similar to that for infants ≥38 weeks.[12]

FIGURE 7. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY GESTATIONAL AGE, HAWAI'I, 1986-2005



There were also differences among broad categories and specific birth defects by gestational age (Table 7).

[12] The gestational age categories for birth defects correspond to the gestational age categories for live births provided by the DOH Office of Health Status Monitoring.

TABLE 7. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY GESTATIONAL AGE, HAWAI‘I, 1986-2005

Diagnosis	<20 weeks N=175		20-27 weeks N=1,969		28-30 weeks N=1,917		31-33 weeks N=4,676		34-37 weeks N=52,442		38-40 weeks N=245,539		>40 weeks N=49,399	
	N	Rate*	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
BRAIN & NERVOUS SYSTEM	0	0.0	61	309.8	65	339.1	92	196.7	379	72.3	538	21.9	78	15.8
Neural tube defects	0	0.0	5	25.4	2	10.4	7	15.0	71	13.5	79	3.2	7	1.4
Anencephaly	0	0.0	2	10.2	2	10.4	1	2.1	10	1.9	5	0.2	3	0.6
Spina bifida	0	0.0	2	10.2	0	0.0	4	8.6	51	9.7	57	2.3	3	0.6
Encephalocele	0	0.0	2	10.2	0	0.0	2	4.3	12	2.3	18	0.7	1	0.2
Microcephaly	0	0.0	11	55.9	15	78.2	16	34.2	133	25.4	159	6.5	26	5.3
Holoprosencephaly	0	0.0	0	0.0	1	5.2	5	10.7	11	2.1	14	0.6	1	0.2
Hydrocephaly (w/o spina bifida)	0	0.0	26	132.0	23	120.0	33	70.6	96	18.3	120	4.9	18	3.6
EYE, EAR, FACE, & NECK	1	57.1	33	167.6	39	203.4	101	216.0	511	97.4	943	38.4	139	28.1
EYE	0	0.0	7	35.6	16	83.5	37	79.1	208	39.7	363	14.8	56	11.3
Anophthalmia	0	0.0	0	0.0	1	5.2	0	0.0	3	0.6	4	0.2	0	0.0
Microphthalmia	0	0.0	1	5.1	2	10.4	9	19.2	44	8.4	40	1.6	10	2.0
Cataract	0	0.0	1	5.1	1	5.2	3	6.4	9	1.7	25	1.0	6	1.2
EAR	1	57.1	28	142.2	22	114.8	79	168.9	345	65.8	677	27.6	101	20.4
Absent auditory canal/ear	0	0.0	0	0.0	0	0.0	3	6.4	15	2.9	24	1.0	3	0.6
Microtia	0	0.0	3	15.2	1	5.2	4	8.6	27	5.1	80	3.3	11	2.2
CARDIAC & CIRCULATORY	3	171.4	393	1,995.9	396	2,065.7	474	1,013.7	1,649	314.4	3,387	137.9	571	115.6
Common truncus	0	0.0	1	5.1	1	5.2	0	0.0	4	0.8	10	0.4	6	1.2
Transposition of the great vessels	0	0.0	0	0.0	7	36.5	4	8.6	33	6.3	89	3.6	17	3.4
Tetralogy of Fallot	0	0.0	2	10.2	3	15.6	11	23.5	33	6.3	74	3.0	9	1.8
Single ventricle	0	0.0	1	5.1	2	10.4	1	2.1	5	1.0	11	0.4	5	1.0
Ventricular septal defect	1	57.1	37	187.9	46	240.0	82	175.4	408	77.8	962	39.2	163	33.0
Atrial septal defect	1	57.1	204	1,036.1	172	897.2	200	427.7	738	140.7	1,177	47.9	188	38.1
Endocardial cushion defect	0	0.0	1	5.1	3	15.6	4	8.6	22	4.2	36	1.5	10	2.0
Pulmonary valve atresia/stenosis	0	0.0	10	50.8	18	93.9	26	55.6	87	16.6	164	6.7	31	6.3
Tricuspid valve atresia/stenosis	0	0.0	1	5.1	0	0.0	2	4.3	15	2.9	35	1.4	4	0.8
Ebstein’s anomaly	0	0.0	0	0.0	2	10.4	0	0.0	3	0.6	11	0.4	0	0.0

* Rate = (number of birth defects)/(number of live births)x10,000.
+ Birth weight >2,499 grams.

TABLE 7. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY GESTATIONAL AGE, HAWAI‘I, 1986-2005

Diagnosis	<20 weeks N=175		20-27 weeks N=1,969		28-30 weeks N=1,917		31-33 weeks N=4,676		34-37 weeks N=52,442		38-40 weeks N=245,539		>40 weeks N=49,399	
	N	Rate*	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Aortic valve atresia/stenosis	0	0.0	2	10.2	1	5.2	2	4.3	16	3.1	25	1.0	5	1.0
Mitral valve atresia/stenosis	0	0.0	0	0.0	0	0.0	3	6.4	12	2.3	21	0.9	2	0.4
Hypoplastic left heart syndrome	0	0.0	1	5.1	1	5.2	3	6.4	11	2.1	29	1.2	2	0.4
Dextrocardia	0	0.0	1	5.1	0	0.0	3	6.4	13	2.5	26	1.1	8	1.6
Patent ductus arteriosus +	2	114.3	1	5.1	4	20.9	27	57.7	684	130.4	1,976	80.5	319	64.6
Coarctation of the aorta	0	0.0	1	5.1	3	15.6	5	10.7	25	4.8	58	2.4	11	2.2
Aortic stenosis	0	0.0	0	0.0	1	5.2	0	0.0	0	0.0	2	0.1	0	0.0
Pulmonary artery atresia/stenosis	0	0.0	55	279.3	90	469.5	118	252.4	210	40.0	209	8.5	38	7.7
Total anomalous pulmonary venous return	0	0.0	0	0.0	0	0.0	2	4.3	6	1.1	25	1.0	4	0.8
Partial anomalous pulmonary venous return	0	0.0	0	0.0	0	0.0	0	0.0	5	1.0	10	0.4	1	0.2
Persistent fetal circulation	0	0.0	3	15.2	5	26.1	7	15.0	44	8.4	109	4.4	45	9.1
RESPIRATORY	0	0.0	38	193.0	32	166.9	82	175.4	251	47.9	400	16.3	72	14.6
Choanal atresia/stenosis	0	0.0	1	5.1	0	0.0	3	6.4	8	1.5	32	1.3	6	1.2
Lung cysts	0	0.0	0	0.0	0	0.0	4	8.6	5	1.0	18	0.7	2	0.4
Hypoplastic lung	0	0.0	22	111.7	18	93.9	42	89.8	60	11.4	35	1.4	7	1.4
OROFACIAL & GASTROINTESTINAL	0	0.0	34	172.7	40	208.7	112	239.5	562	107.2	1,148	46.8	198	40.1
Oral clefts	0	0.0	5	25.4	8	41.7	28	59.9	153	29.2	406	16.5	68	13.8
Cleft palate (isolated)	0	0.0	3	15.2	1	5.2	11	23.5	65	12.4	154	6.3	23	4.7
Cleft lip (+/-cleft palate)	0	0.0	2	10.2	7	36.5	17	36.4	88	16.8	252	10.3	45	9.1
TE fistula/Esophageal atresia	0	0.0	1	5.1	1	5.2	12	25.7	22	4.2	27	1.1	11	2.2
Pyloric stenosis	0	0.0	1	5.1	0	0.0	6	12.8	56	10.7	183	7.5	29	5.9
Atresia/stenosis small intestine	0	0.0	2	10.2	3	15.6	14	29.9	49	9.3	31	1.3	2	0.4
Anal atresia/stenosis	0	0.0	1	5.1	9	46.9	15	32.1	55	10.5	81	3.3	10	2.0
Hirschsprung’s disease	0	0.0	3	15.2	0	0.0	3	6.4	22	4.2	54	2.2	11	2.2
Malrotation	0	0.0	1	5.1	1	5.2	8	17.1	33	6.3	46	1.9	4	0.8
Biliary atresia	0	0.0	0	0.0	2	10.4	1	2.1	7	1.3	26	1.1	3	0.6

* Rate = (number of birth defects)/(number of live births)x10,000.
\+ Birth weight >2,499 grams.

TABLE 7. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY GESTATIONAL AGE, HAWAI‘I, 1986-2005

Diagnosis	<20 weeks N=175		20-27 weeks N=1,969		28-30 weeks N=1,917		31-33 weeks N=4,676		34-37 weeks N=52,442		38-40 weeks N=245,539		>40 weeks N=49,399	
	N	Rate*	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
GENITAL & URINARY	1	57.1	111	563.7	109	568.6	188	402.1	911	173.7	2,280	92.9	342	69.2
Ovarian cyst	0	0.0	1	5.1	1	5.2	2	4.3	1	0.2	9	0.4	5	1.0
Undescended testicle +	0	0.0	0	0.0	0	0.0	8	17.1	223	42.5	794	32.3	142	28.7
Hypospadias/Epispadias/Chordee	1	57.1	14	71.1	26	135.6	67	143.3	264	50.3	767	31.2	100	20.2
Ambiguous genitalia	0	0.0	2	10.2	0	0.0	0	0.0	2	0.4	8	0.3	2	0.4
Absent/hypoplastic kidney	0	0.0	4	20.3	8	41.7	20	42.8	50	9.5	49	2.0	9	1.8
Cystic kidney	0	0.0	4	20.3	8	41.7	18	38.5	45	8.6	64	2.6	6	1.2
Hydronephrosis	0	0.0	21	106.7	21	109.5	35	74.9	151	28.8	242	9.9	22	4.5
Exstrophy bladder	0	0.0	0	0.0	0	0.0	0	0.0	3	0.6	4	0.2	1	0.2
Posterior urethral valves	0	0.0	0	0.0	1	5.2	2	4.3	5	1.0	8	0.3	1	0.2
LIMB & MUSCULOSKELETAL	1	57.1	51	259.0	77	401.7	163	348.6	1,084	206.7	2,755	112.2	517	104.7
Scoliosis	0	0.0	4	20.3	3	15.6	6	12.8	26	5.0	29	1.2	6	1.2
Hip dislocation	0	0.0	2	10.2	3	15.6	8	17.1	48	9.2	233	9.5	77	15.6
Clubfoot	1	57.1	11	55.9	15	78.2	28	59.9	251	47.9	731	29.8	117	23.7
Polydactyly	0	0.0	1	5.1	5	26.1	20	42.8	104	19.8	419	17.1	69	14.0
Syndactyly	0	0.0	5	25.4	5	26.1	18	38.5	68	13.0	147	6.0	39	7.9
Limb reduction deformity - upper	0	0.0	9	45.7	2	10.4	7	15.0	31	5.9	49	2.0	14	2.8
Limb reduction deformity - lower	0	0.0	6	30.5	0	0.0	3	6.4	14	2.7	19	0.8	5	1.0
Craniosynostosis	0	0.0	1	5.1	1	5.2	3	6.4	45	8.6	98	4.0	25	5.1
Crouzon’s disease	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	2	0.1	0	0.0
Apert syndrome	0	0.0	0	0.0	0	0.0	0	0.0	5	1.0	6	0.2	0	0.0
Goldenhar syndrome	0	0.0	1	5.1	0	0.0	1	2.1	6	1.1	12	0.5	1	0.2
Spina bifida occulta	0	0.0	0	0.0	1	5.2	0	0.0	1	0.2	7	0.3	1	0.2
Dwarfism	0	0.0	3	15.2	4	20.9	4	8.6	26	5.0	42	1.7	9	1.8
Osteogenesis imperfecta	0	0.0	0	0.0	1	5.2	0	0.0	5	1.0	6	0.2	2	0.4
Diaphragmatic hernia	0	0.0	3	15.2	2	10.4	6	12.8	17	3.2	40	1.6	8	1.6
Abdominal wall defects	0	0.0	5	25.4	7	36.5	18	38.5	128	24.4	78	3.2	14	2.8

* Rate = (number of birth defects)/(number of live births)x10,000.
+ Birth weight >2,499 grams.

TABLE 7. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY GESTATIONAL AGE, HAWAI'I, 1986-2005

Diagnosis	<20 weeks N=175		20-27 weeks N=1,969		28-30 weeks N=1,917		31-33 weeks N=4,676		34-37 weeks N=52,442		38-40 weeks N=245,539		>40 weeks N=49,399	
	N	Rate*	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Omphalocele	0	0.0	2	10.2	3	15.6	5	10.7	27	5.1	17	0.7	4	0.8
Gastroschisis	0	0.0	1	5.1	1	5.2	10	21.4	83	15.8	17	0.7	2	0.4
Prune-belly syndrome	0	0.0	1	5.1	2	10.4	1	2.1	4	0.8	1	0.0	0	0.0
Poland syndrome	0	0.0	0	0.0	0	0.0	1	2.1	1	0.2	4	0.2	0	0.0
SKIN & INTEGUMENT	1	57.1	10	50.8	21	109.5	56	119.8	595	113.5	1,370	55.8	211	42.7
CHROMOSOME & SYNDROME	2	114.3	19	96.5	24	125.2	64	136.9	378	72.1	651	26.5	92	18.6
Down syndrome (trisomy 21)	1	57.1	1	5.1	4	20.9	10	21.4	128	24.4	171	7.0	12	2.4
Patau syndrome (trisomy 13)	0	0.0	0	0.0	2	10.4	2	4.3	15	2.9	7	0.3	2	0.4
Edwards syndrome (trisomy 18)	0	0.0	3	15.2	3	15.6	9	19.2	17	3.2	17	0.7	5	1.0
Cri du Chat syndrome	0	0.0	0	0.0	0	0.0	0	0.0	2	0.4	1	0.0	2	0.4
Wolff-Hirschorn syndrome	0	0.0	0	0.0	0	0.0	1	2.1	4	0.8	2	0.1	0	0.0
Autosomal anomalies in normal infant	0	0.0	1	5.1	2	10.4	2	4.3	23	4.4	90	3.7	11	2.2
Triploidy	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Turner syndrome (45,X)	1	57.1	1	5.1	0	0.0	0	0.0	8	1.5	26	1.1	1	0.2
Klinefelter syndrome (47,XXY)	0	0.0	0	0.0	0	0.0	0	0.0	8	1.5	26	1.1	1	0.2
Absent spleen/Asplenia	0	0.0	0	0.0	0	0.0	1	2.1	3	0.6	4	0.2	0	0.0
Accessory spleen	0	0.0	1	5.1	3	15.6	2	4.3	12	2.3	9	0.4	2	0.4
Situs inversus	0	0.0	0	0.0	0	0.0	2	4.3	10	1.9	23	0.9	5	1.0
Conjoined twins	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	2	0.1	0	0.0
Tuberous sclerosis	0	0.0	0	0.0	0	0.0	1	2.1	3	0.6	10	0.4	1	0.2
Amniotic band syndrome	0	0.0	6	30.5	3	15.6	4	8.6	20	3.8	22	0.9	7	1.4
DiGeorge syndrome	0	0.0	0	0.0	0	0.0	0	0.0	5	1.0	17	0.7	3	0.6
Pierre-Robin sequence	0	0.0	0	0.0	0	0.0	2	4.3	7	1.3	24	1.0	2	0.4
Werdnig-Hoffman syndrome	0	0.0	1	5.1	2	10.4	2	4.3	2	0.4	5	0.2	1	0.2
Wolff-Parkinson-White syndrome	0	0.0	0	0.0	1	5.2	0	0.0	1	0.2	15	0.6	2	0.4
NEOPLASMS	0	0.0	4	20.3	2	10.4	7	15.0	49	9.3	142	5.8	21	4.3
TOTAL BIRTH DEFECT CASES	5	285.7	443	2,249.9	476	2,483.0	700	1,497.0	3,454	658.6	9,126	371.7	1,494	302.4

* Rate = (number of birth defects)/(number of live births)x10,000.
\+ Birth weight >2,499 grams.

TABLE 7. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY GESTATIONAL AGE, HAWAI‘I, 1986-2005

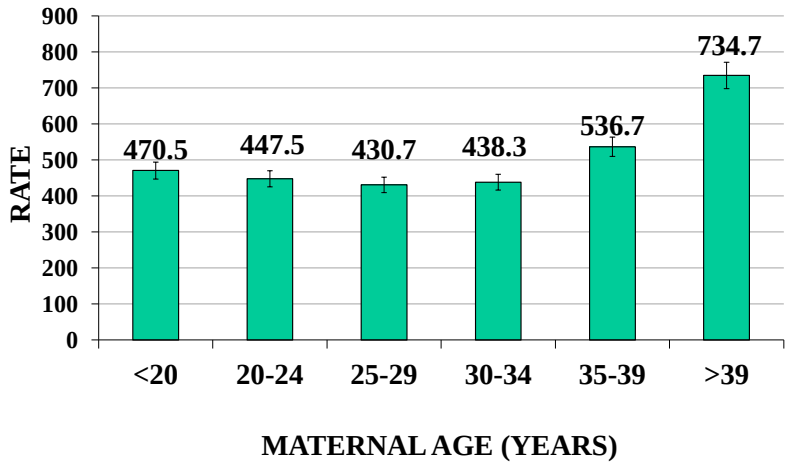
Diagnosis	<20 weeks N=175		20-27 weeks N=1,969		28-30 weeks N=1,917		31-33 weeks N=4,676		34-37 weeks N=52,442		38-40 weeks N=245,539		>40 weeks N=49,399	
	N	Rate*	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
FETAL ALCOHOL SYNDROME	0	0.0	2	10.2	4	20.9	3	6.4	22	4.2	46	1.9	9	1.8
BIRTH DEFECTS CASES WITH:														
CONGENITAL INFECTION	0	0.0	19	96.5	22	114.8	16	34.2	66	12.6	109	4.4	18	3.6
MATERNAL ALCOHOL USE	0	0.0	42	213.3	51	266.0	57	121.9	326	62.2	774	31.5	144	29.2
MATERNAL TOBACCO USE	1	57.1	122	619.6	142	740.7	217	464.1	1,224	233.4	2,666	108.6	394	79.8
MATERNAL ILLICIT DRUG USE	2	114.3	59	299.6	67	349.5	136	290.8	918	175.1	1,693	69.0	198	40.1
Marijuana	0	0.0	23	116.8	19	99.1	54	115.5	241	46.0	536	21.8	97	19.6
Cocaine	0	0.0	13	66.0	12	62.6	32	68.4	169	32.2	310	12.6	27	5.5
Methamphetamine/Amphetamine	2	114.3	32	162.5	42	219.1	91	194.6	685	130.6	1,109	45.2	109	22.1

* Rate = (number of birth defects)/(number of live births)x10,000.
+ Birth weight >2,499 grams.

Birth Defect Rates by Maternal Age

Birth defect rates were similar for maternal age groups up to age 34 years, with higher rates for older mothers age 35-39 years (536.7 per 10,000 live births) and age >39 years (734.7) (Figure 8).

FIGURE 8. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY MATERNAL AGE, HAWAI'I, 1986-2005



There were also differences among broad categories and specific birth defects by maternal age (Table 8).

TABLE 8. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY MATERNAL AGE, HAWAI‘I, 1986-2005

Diagnosis	<20 years N=36,218		20-24 years N=95,571		25-29 years N=101,985		30-34 years N=84,883		35-39 years N=42,593		>39 years N=9,160	
	N	Rate*	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
BRAIN & NERVOUS SYSTEM	194	53.6	393	41.1	389	38.1	307	36.2	193	45.3	59	64.4
Neural tube defects	42	11.6	75	7.8	110	10.8	74	8.7	44	10.3	12	13.1
Anencephaly	21	5.8	23	2.4	45	4.4	30	3.5	11	2.6	2	2.2
Spina bifida	18	5.0	34	3.6	41	4.0	35	4.1	28	6.6	6	6.6
Encephalocele	3	0.8	20	2.1	28	2.7	11	1.3	7	1.6	4	4.4
Microcephaly	48	13.3	110	11.5	72	7.1	76	9.0	51	12.0	10	10.9
Holoprosencephaly	9	2.5	14	1.5	7	0.7	10	1.2	4	0.9	3	3.3
Hydrocephaly (w/o spina bifida)	45	12.4	106	11.1	125	12.3	73	8.6	43	10.1	11	12.0
EYE, EAR, FACE, & NECK	181	50.0	455	47.6	453	44.4	419	49.4	269	63.2	114	124.5
EYE	66	18.2	185	19.4	171	16.8	135	15.9	101	23.7	51	55.7
Anophthalmia	2	0.6	4	0.4	5	0.5	3	0.4	0	0.0	0	0.0
Microphthalmia	13	3.6	23	2.4	31	3.0	23	2.7	15	3.5	3	3.3
Cataract	6	1.7	14	1.5	14	1.4	6	0.7	3	0.7	2	2.2
EAR	132	36.4	318	33.3	321	31.5	303	35.7	186	43.7	71	77.5
Absent auditory canal/ear	7	1.9	14	1.5	10	1.0	9	1.1	4	0.9	1	1.1
Microtia	11	3.0	34	3.6	27	2.6	33	3.9	21	4.9	4	4.4
CARDIAC & CIRCULATORY	749	206.8	1,742	182.3	1,811	177.6	1,585	186.7	905	212.5	262	286.0
Common truncus	1	0.3	7	0.7	6	0.6	5	0.6	7	1.6	1	1.1
Transposition of the great vessels	13	3.6	43	4.5	44	4.3	38	4.5	17	4.0	7	7.6
Tetralogy of Fallot	16	4.4	38	4.0	37	3.6	28	3.3	15	3.5	6	6.6
Single ventricle	5	1.4	7	0.7	6	0.6	5	0.6	4	0.9	1	1.1
Ventricular septal defect	151	41.7	427	44.7	445	43.6	414	48.8	240	56.3	74	80.8
Atrial septal defect	300	82.8	631	66.0	687	67.4	598	70.4	378	88.7	131	143.0
Endocardial cushion defect	9	2.5	16	1.7	22	2.2	13	1.5	21	4.9	6	6.6
Pulmonary valve atresia/stenosis	43	11.9	92	9.6	85	8.3	70	8.2	43	10.1	14	15.3
Tricuspid valve atresia/stenosis	10	2.8	16	1.7	12	1.2	12	1.4	6	1.4	3	3.3
Ebstein’s anomaly	0	0.0	3	0.3	8	0.8	5	0.6	2	0.5	1	1.1

* Rate = (number of birth defects)/(number of live births)x10,000.
+ Birth weight >2,499 grams.

TABLE 8. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY MATERNAL AGE, HAWAI‘I, 1986-2005

Diagnosis	<20 years N=36,218		20-24 years N=95,571		25-29 years N=101,985		30-34 years N=84,883		35-39 years N=42,593		>39 years N=9,160	
	N	Rate*	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Aortic valve atresia/stenosis	5	1.4	9	0.9	15	1.5	13	1.5	10	2.3	1	1.1
Mitral valve atresia/stenosis	3	0.8	11	1.2	9	0.9	3	0.4	10	2.3	2	2.2
Hypoplastic left heart syndrome	6	1.7	17	1.8	13	1.3	8	0.9	9	2.1	5	5.5
Dextrocardia	3	0.8	15	1.6	18	1.8	7	0.8	8	1.9	2	2.2
Patent ductus arteriosus +	293	80.9	738	77.2	815	79.9	681	80.2	372	87.3	119	129.9
Coarctation of the aorta	10	2.8	26	2.7	35	3.4	20	2.4	11	2.6	2	2.2
Aortic stenosis	0	0.0	0	0.0	0	0.0	1	0.1	1	0.2	1	1.1
Pulmonary artery atresia/stenosis	92	25.4	168	17.6	189	18.5	145	17.1	98	23.0	32	34.9
Total anomalous pulmonary venous return	2	0.6	4	0.4	13	1.3	12	1.4	6	1.4	0	0.0
Partial anomalous pulmonary venous return	1	0.3	5	0.5	4	0.4	7	0.8	0	0.0	0	0.0
Persistent fetal circulation	9	2.5	55	5.8	56	5.5	58	6.8	30	7.0	5	5.5
RESPIRATORY	105	29.0	260	27.2	244	23.9	177	20.9	107	25.1	45	49.1
Choanal atresia/stenosis	3	0.8	13	1.4	11	1.1	13	1.5	8	1.9	2	2.2
Lung cysts	6	1.7	10	1.0	8	0.8	6	0.7	1	0.2	0	0.0
Hypoplastic lung	29	8.0	59	6.2	62	6.1	35	4.1	23	5.4	4	4.4
OROFACIAL & GASTROINTESTINAL	229	63.2	563	58.9	603	59.1	478	56.3	256	60.1	87	95.0
Oral clefts	66	18.2	186	19.5	199	19.5	162	19.1	88	20.7	31	33.8
Cleft palate (isolated)	27	7.5	64	6.7	67	6.6	67	7.9	39	9.2	11	12.0
Cleft lip (+/-cleft palate)	39	10.8	123	12.9	132	12.9	95	11.2	49	11.5	20	21.8
TE fistula/Esophageal atresia	9	2.5	14	1.5	27	2.6	21	2.5	3	0.7	4	4.4
Pyloric stenosis	43	11.9	70	7.3	68	6.7	68	8.0	27	6.3	7	7.6
Atresia/stenosis small intestine	10	2.8	28	2.9	27	2.6	20	2.4	14	3.3	8	8.7
Anal atresia/stenosis	21	5.8	39	4.1	56	5.5	40	4.7	23	5.4	5	5.5
Hirschsprung’s disease	15	4.1	20	2.1	28	2.7	17	2.0	11	2.6	1	1.1
Malrotation	12	3.3	35	3.7	29	2.8	19	2.2	3	0.7	3	3.3
Biliary atresia	4	1.1	14	1.5	11	1.1	8	0.9	2	0.5	0	0.0
GENITAL & URINARY	425	117.3	1,117	116.9	1,087	106.6	854	100.6	474	111.3	117	127.7

* Rate = (number of birth defects)/(number of live births)x10,000.
+ Birth weight >2,499 grams.

TABLE 8. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY MATERNAL AGE, HAWAI‘I, 1986-2005

Diagnosis	<20 years N=36,218		20-24 years N=95,571		25-29 years N=101,985		30-34 years N=84,883		35-39 years N=42,593		>39 years N=9,160	
	N	Rate*	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Ovarian cyst	1	0.3	6	0.6	5	0.5	4	0.5	3	0.7	0	0.0
Undescended testicle +	141	38.9	351	36.7	302	29.6	244	28.7	112	26.3	19	20.7
Hypospadias/Epispadias/Chordee	128	35.3	330	34.5	342	33.5	248	29.2	167	39.2	37	40.4
Ambiguous genitalia	1	0.3	7	0.7	3	0.3	5	0.6	4	0.9	0	0.0
Absent/hypoplastic kidney	18	5.0	45	4.7	57	5.6	32	3.8	15	3.5	6	6.6
Cystic kidney	14	3.9	45	4.7	44	4.3	33	3.9	20	4.7	9	9.8
Hydronephrosis	48	13.3	136	14.2	139	13.6	112	13.2	59	13.9	17	18.6
Exstrophy bladder	0	0.0	1	0.1	2	0.2	4	0.5	1	0.2	1	1.1
Posterior urethral valves	2	0.6	9	0.9	4	0.4	6	0.7	1	0.2	1	1.1
LIMB & MUSCULOSKELETAL	519	143.3	1,309	137.0	1,305	128.0	1,053	124.1	556	130.5	172	187.8
Scoliosis	17	4.7	21	2.2	25	2.5	20	2.4	8	1.9	3	3.3
Hip dislocation	36	9.9	77	8.1	104	10.2	90	10.6	58	13.6	9	9.8
Clubfoot	147	40.6	378	39.6	310	30.4	241	28.4	100	23.5	27	29.5
Polydactyly	62	17.1	190	19.9	183	17.9	141	16.6	43	10.1	22	24.0
Syndactyly	31	8.6	84	8.8	78	7.6	62	7.3	28	6.6	14	15.3
Limb reduction deformity - upper	19	5.2	33	3.5	30	2.9	31	3.7	12	2.8	4	4.4
Limb reduction deformity - lower	5	1.4	20	2.1	12	1.2	11	1.3	4	0.9	2	2.2
Craniosynostosis	12	3.3	39	4.1	60	5.9	36	4.2	27	6.3	4	4.4
Crouzon’s disease	0	0.0	1	0.1	1	0.1	0	0.0	0	0.0	0	0.0
Apert syndrome	0	0.0	2	0.2	1	0.1	1	0.1	6	1.4	1	1.1
Goldenhar syndrome	3	0.8	3	0.3	5	0.5	3	0.4	6	1.4	1	1.1
Spina bifida occulta	1	0.3	1	0.1	5	0.5	2	0.2	2	0.5	2	2.2
Dwarfism	10	2.8	22	2.3	24	2.4	23	2.7	19	4.5	4	4.4
Osteogenesis imperfecta	3	0.8	6	0.6	2	0.2	4	0.5	0	0.0	0	0.0
Diaphragmatic hernia	5	1.4	21	2.2	27	2.6	17	2.0	14	3.3	4	4.4
Abdominal wall defects	52	14.4	107	11.2	69	6.8	54	6.4	27	6.3	12	13.1
Omphalocele	6	1.7	27	2.8	17	1.7	26	3.1	14	3.3	10	10.9

* Rate = (number of birth defects)/(number of live births)x10,000.
+ Birth weight >2,499 grams.

TABLE 8. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY MATERNAL AGE, HAWAI‘I, 1986-2005

Diagnosis	<20 years N=36,218		20-24 years N=95,571		25-29 years N=101,985		30-34 years N=84,883		35-39 years N=42,593		>39 years N=9,160	
	N	Rate*	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Gastroschisis	41	11.3	55	5.8	23	2.3	9	1.1	3	0.7	1	1.1
Prune-belly syndrome	1	0.3	4	0.4	2	0.2	2	0.2	1	0.2	0	0.0
Poland syndrome	1	0.3	1	0.1	3	0.3	1	0.1	0	0.0	0	0.0
SKIN & INTEGUMENT	238	65.7	613	64.1	602	59.0	479	56.4	272	63.9	86	93.9
CHROMOSOME & SYNDROME	116	32.0	311	32.5	354	34.7	431	50.8	554	130.1	294	321.0
Down syndrome (trisomy 21)	19	5.2	63	6.6	71	7.0	110	13.0	173	40.6	129	140.8
Patau syndrome (trisomy 13)	2	0.6	8	0.8	7	0.7	19	2.2	23	5.4	14	15.3
Edwards syndrome (trisomy 18)	6	1.7	18	1.9	29	2.8	35	4.1	38	8.9	42	45.9
Cri du Chat syndrome	0	0.0	0	0.0	2	0.2	3	0.4	3	0.7	0	0.0
Wolff-Hirschorn syndrome	1	0.3	3	0.3	0	0.0	3	0.4	0	0.0	1	1.1
Autosomal anomalies in normal infant	3	0.8	6	0.6	11	1.1	40	4.7	66	15.5	14	15.3
Triploidy	2	0.6	7	0.7	4	0.4	11	1.3	9	2.1	6	6.6
Turner syndrome (45,X)	10	2.8	30	3.1	31	3.0	27	3.2	38	8.9	11	12.0
Klinefelter syndrome (47,XXY)	0	0.0	7	0.7	6	0.6	6	0.7	25	5.9	10	10.9
Absent spleen/Asplenia	1	0.3	5	0.5	3	0.3	2	0.2	2	0.5	0	0.0
Accessory spleen	9	2.5	11	1.2	9	0.9	8	0.9	4	0.9	1	1.1
Situs inversus	3	0.8	10	1.0	16	1.6	9	1.1	5	1.2	0	0.0
Conjoined twins	0	0.0	7	0.7	2	0.2	2	0.2	2	0.5	0	0.0
Tuberous sclerosis	0	0.0	5	0.5	4	0.4	5	0.6	1	0.2	0	0.0
Amniotic band syndrome	12	3.3	25	2.6	20	2.0	20	2.4	4	0.9	1	1.1
DiGeorge syndrome	1	0.3	9	0.9	7	0.7	5	0.6	3	0.7	0	0.0
Pierre-Robin sequence	7	1.9	8	0.8	13	1.3	6	0.7	2	0.5	0	0.0
Werdnig-Hoffman syndrome	1	0.3	4	0.4	6	0.6	3	0.4	1	0.2	1	1.1
Wolff-Parkinson-White syndrome	6	1.7	5	0.5	3	0.3	5	0.6	1	0.2	0	0.0
NEOPLASMS	31	8.6	90	9.4	94	9.2	102	12.0	42	9.9	10	10.9
TOTAL BIRTH DEFECT CASES	1,704	470.5	4,277	447.5	4,393	430.7	3,720	438.3	2,286	536.7	673	734.7

* Rate = (number of birth defects)/(number of live births)x10,000.
\+ Birth weight >2,499 grams.

TABLE 8. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY MATERNAL AGE, HAWAI‘I, 1986-2005

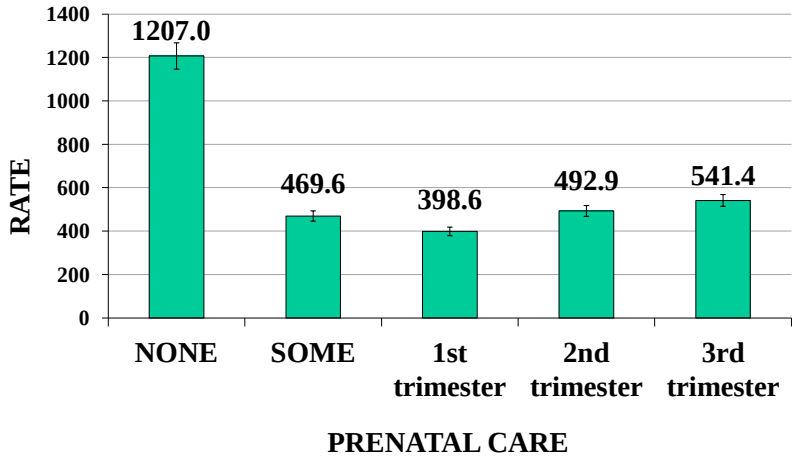
Diagnosis	<20 years N=36,218		20-24 years N=95,571		25-29 years N=101,985		30-34 years N=84,883		35-39 years N=42,593		>39 years N=9,160	
	N	Rate*	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
FETAL ALCOHOL SYNDROME	11	3.0	24	2.5	17	1.7	17	2.0	18	4.2	3	3.3
BIRTH DEFECTS CASES WITH:												
CONGENITAL INFECTION	42	11.6	93	9.7	50	4.9	43	5.1	31	7.3	9	9.8
MATERNAL ALCOHOL USE	159	43.9	368	38.5	380	37.3	341	40.2	228	53.5	52	56.8
MATERNAL TOBACCO USE	693	191.3	1,482	155.1	1,287	126.2	968	114.0	476	111.8	93	101.5
MATERNAL ILLICIT DRUG USE	413	114.0	919	96.2	878	86.1	617	72.7	295	69.3	52	56.8
Marijuana	199	54.9	304	31.8	239	23.4	165	19.4	85	20.0	16	17.5
Cocaine	53	14.6	141	14.8	176	17.3	134	15.8	77	18.1	9	9.8
Methamphetamine/Amphetamine	226	62.4	644	67.4	615	60.3	422	49.7	188	44.1	29	31.7

* Rate = (number of birth defects)/(number of live births)x10,000.
+ Birth weight >2,499 grams.

Birth Defect Rates by Prenatal Care

The birth defect rate for mothers who received no prenatal care (1,207.0 per 10,000 live births) was over 2.5 times higher than the rate for mothers who received some prenatal care (469.6) (Figure 9). There was a gradual increase in birth defect rate with later entry into prenatal care. Those with prenatal care beginning in the third trimester had a rate (541.4) that was higher than those with first trimester (398.6) and second trimester (492.9) entry.

FIGURE 9. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY PRENATAL CARE, HAWAI'I , 1986-2005



There were also differences among broad categories and specific birth defects for birth defect rates by prenatal care (Table 9).

TABLE 9. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY PRENATAL CARE, HAWAI‘I, 1986-2005

Diagnosis	None N=2,676		Yes-ever N=350,297		Yes-1st Trimester N=283,856		Yes-2nd Trimester N=54,085		Yes-3rd Trimester N=12,356	
	N	Rate*	N	Rate	N	Rate	N	Rate	N	Rate
BRAIN & NERVOUS SYSTEM	43	160.7	1,473	42.1	890	31.4	262	48.4	62	50.2
Neural tube defects	2	7.5	349	10.0	192	6.8	38	7.0	10	8.1
Anencephaly	2	7.5	129	3.7	63	2.2	14	2.6	1	0.8
Spina bifida	0	0.0	158	4.5	97	3.4	17	3.1	7	5.7
Encephalocele	0	0.0	72	2.1	38	1.3	7	1.3	2	1.6
Microcephaly	17	63.5	346	9.9	209	7.4	87	16.1	18	14.6
Holoprosencephaly	1	3.7	47	1.3	19	0.7	13	2.4	3	2.4
Hydrocephaly (w/o spina bifida)	12	44.8	384	11.0	232	8.2	67	12.4	20	16.2
EYE, EAR, FACE, & NECK	53	198.1	1,823	52.0	1,229	43.3	344	63.6	84	68.0
EYE	17	63.5	685	19.6	469	16.5	126	23.3	39	31.6
Anophthalmia	1	3.7	13	0.4	7	0.2	3	0.6	1	0.8
Microphthalmia	3	11.2	104	3.0	72	2.5	17	3.1	7	5.7
Cataract	0	0.0	45	1.3	35	1.2	6	1.1	3	2.4
EAR	38	142.0	1,282	36.6	862	30.4	253	46.8	57	46.1
Absent auditory canal/ear	3	11.2	42	1.2	29	1.0	4	0.7	5	4.0
Microtia	1	3.7	128	3.7	85	3.0	33	6.1	3	2.4
CARDIAC & CIRCULATORY	180	672.6	6,787	193.7	4,763	167.8	1,139	210.6	293	237.1
Common truncus	0	0.0	27	0.8	22	0.8	1	0.2	0	0.0
Transposition of the great vessels	7	26.2	154	4.4	93	3.3	30	5.5	12	9.7
Tetralogy of Fallot	2	7.5	137	3.9	99	3.5	19	3.5	7	5.7
Single ventricle	2	7.5	26	0.7	15	0.5	8	1.5	1	0.8
Ventricular septal defect	42	157.0	1,688	48.2	1,185	41.7	285	52.7	74	59.9
Atrial septal defect	75	280.3	2,611	74.5	1,836	64.7	454	83.9	125	101.2
Endocardial cushion defect	3	11.2	84	2.4	57	2.0	12	2.2	2	1.6
Pulmonary valve atresia/stenosis	6	22.4	333	9.5	232	8.2	47	8.7	17	13.8
Tricuspid valve atresia/stenosis	1	3.7	58	1.7	37	1.3	10	1.8	6	4.9
Ebstein’s anomaly	0	0.0	17	0.5	10	0.4	3	0.6	1	0.8

* Rate = (number of birth defects)/(number of live births)x10,000.
+ Birth weight >2,499 grams.

TABLE 9. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY PRENATAL CARE, HAWAI'I, 1986-2005

Diagnosis	None N=2,676		Yes-ever N=350,297		Yes-1st Trimester N=283,856		Yes-2nd Trimester N=54,085		Yes-3rd Trimester N=12,356	
	N	Rate*	N	Rate	N	Rate	N	Rate	N	Rate
Aortic valve atresia/stenosis	0	0.0	50	1.4	32	1.1	13	2.4	1	0.8
Mitral valve atresia/stenosis	3	11.2	35	1.0	26	0.9	6	1.1	2	1.6
Hypoplastic left heart syndrome	1	3.7	57	1.6	27	1.0	14	2.6	3	2.4
Dextrocardia	2	7.5	50	1.4	30	1.1	10	1.8	6	4.9
Patent ductus arteriosus +	39	145.7	2,956	84.4	2,146	75.6	484	89.5	137	110.9
Coarctation of the aorta	0	0.0	102	2.9	74	2.6	18	3.3	1	0.8
Aortic stenosis	0	0.0	3	0.1	2	0.1	0	0.0	0	0.0
Pulmonary artery atresia/stenosis	23	85.9	694	19.8	497	17.5	119	22.0	23	18.6
Total anomalous pulmonary venous return	1	3.7	36	1.0	27	1.0	5	0.9	3	2.4
Partial anomalous pulmonary venous return	0	0.0	17	0.5	12	0.4	4	0.7	0	0.0
Persistent fetal circulation	6	22.4	202	5.8	128	4.5	48	8.9	11	8.9
RESPIRATORY	20	74.7	908	25.9	610	21.5	176	32.5	37	29.9
Choanal atresia/stenosis	2	7.5	46	1.3	33	1.2	11	2.0	0	0.0
Lung cysts	0	0.0	30	0.9	24	0.8	5	0.9	1	0.8
Hypoplastic lung	4	14.9	206	5.9	127	4.5	43	8.0	6	4.9
OROFACIAL & GASTROINTESTINAL	42	157.0	2,150	61.4	1,476	52.0	376	69.5	97	78.5
Oral clefts	13	48.6	713	20.4	499	17.6	125	23.1	27	21.9
Cleft palate (isolated)	7	26.2	266	7.6	187	6.6	52	9.6	9	7.3
Cleft lip (+/-cleft palate)	6	22.4	448	12.8	312	11.0	74	13.7	18	14.6
TE fistula/Esophageal atresia	0	0.0	78	2.2	55	1.9	11	2.0	5	4.0
Pyloric stenosis	2	7.5	272	7.8	187	6.6	48	8.9	6	4.9
Atresia/stenosis small intestine	2	7.5	106	3.0	74	2.6	12	2.2	1	0.8
Anal atresia/stenosis	1	3.7	183	5.2	130	4.6	33	6.1	8	6.5
Hirschsprung's disease	2	7.5	88	2.5	63	2.2	15	2.8	3	2.4
Malrotation	0	0.0	100	2.9	65	2.3	19	3.5	5	4.0
Biliary atresia	0	0.0	39	1.1	19	0.7	15	2.8	1	0.8
GENITAL & URINARY	66	246.6	3,948	112.7	2,743	96.6	701	129.6	200	161.9

* Rate = (number of birth defects)/(number of live births)x10,000.
+ Birth weight >2,499 grams.

TABLE 9. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY PRENATAL CARE, HAWAI‘I, 1986-2005

Diagnosis	None N=2,676		Yes-ever N=350,297		Yes-1st Trimester N=283,856		Yes-2nd Trimester N=54,085		Yes-3rd Trimester N=12,356	
	N	Rate*	N	Rate	N	Rate	N	Rate	N	Rate
Ovarian cyst	0	0.0	19	0.5	11	0.4	4	0.7	1	0.8
Undescended testicle +	11	41.1	1,143	32.6	771	27.2	251	46.4	54	43.7
Hypospadias/Epispadias/Chordee	15	56.1	1,224	34.9	888	31.3	207	38.3	60	48.6
Ambiguous genitalia	1	3.7	19	0.5	12	0.4	3	0.6	2	1.6
Absent/hypoplastic kidney	4	14.9	168	4.8	102	3.6	22	4.1	12	9.7
Cystic kidney	1	3.7	160	4.6	110	3.9	24	4.4	10	8.1
Hydronephrosis	5	18.7	504	14.4	354	12.5	69	12.8	24	19.4
Exstrophy bladder	0	0.0	8	0.2	5	0.2	1	0.2	1	0.8
Posterior urethral valves	1	3.7	22	0.6	16	0.6	1	0.2	0	0.0
LIMB & MUSCULOSKELETAL	86	321.4	4,760	135.9	3,301	116.3	789	145.9	204	165.1
Scoliosis	1	3.7	92	2.6	53	1.9	19	3.5	0	0.0
Hip dislocation	3	11.2	365	10.4	279	9.8	56	10.4	13	10.5
Clubfoot	20	74.7	1,159	33.1	775	27.3	190	35.1	63	51.0
Polydactyly	9	33.6	629	18.0	454	16.0	105	19.4	21	17.0
Syndactyly	5	18.7	291	8.3	202	7.1	48	8.9	15	12.1
Limb reduction deformity - upper	4	14.9	124	3.5	79	2.8	26	4.8	5	4.0
Limb reduction deformity - lower	0	0.0	53	1.5	36	1.3	9	1.7	2	1.6
Craniosynostosis	2	7.5	168	4.8	125	4.4	20	3.7	6	4.9
Crouzon’s disease	0	0.0	2	0.1	1	0.0	0	0.0	1	0.8
Apert syndrome	0	0.0	11	0.3	9	0.3	2	0.4	0	0.0
Goldenhar syndrome	0	0.0	21	0.6	16	0.6	3	0.6	1	0.8
Spina bifida occulta	0	0.0	11	0.3	8	0.3	0	0.0	1	0.8
Dwarfism	1	3.7	101	2.9	69	2.4	13	2.4	5	4.0
Osteogenesis imperfecta	0	0.0	15	0.4	11	0.4	3	0.6	0	0.0
Diaphragmatic hernia	1	3.7	87	2.5	53	1.9	20	3.7	1	0.8
Abdominal wall defects	6	22.4	312	8.9	180	6.3	53	9.8	9	7.3
Omphalocele	0	0.0	99	2.8	46	1.6	17	3.1	2	1.6

* Rate = (number of birth defects)/(number of live births)x10,000.
+ Birth weight >2,499 grams.

TABLE 9. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY PRENATAL CARE, HAWAI'I, 1986-2005

Diagnosis	None N=2,676		Yes-ever N=350,297		Yes-1st Trimester N=283,856		Yes-2nd Trimester N=54,085		Yes-3rd Trimester N=12,356	
	N	Rate*	N	Rate	N	Rate	N	Rate	N	Rate
Gastroschisis	5	18.7	125	3.6	78	2.7	24	4.4	4	3.2
Prune-belly syndrome	0	0.0	10	0.3	7	0.2	1	0.2	0	0.0
Poland syndrome	0	0.0	6	0.2	5	0.2	0	0.0	1	0.8
SKIN & INTEGUMENT	68	254.1	2,195	62.7	1,488	52.4	442	81.7	141	114.1
CHROMOSOME & SYNDROME	26	97.2	1,966	56.1	1,144	40.3	214	39.6	51	41.3
Down syndrome (trisomy 21)	4	14.9	548	15.6	290	10.2	69	12.8	12	9.7
Patau syndrome (trisomy 13)	3	11.2	69	2.0	35	1.2	4	0.7	3	2.4
Edwards syndrome (trisomy 18)	1	3.7	164	4.7	65	2.3	11	2.0	5	4.0
Cri du Chat syndrome	0	0.0	8	0.2	4	0.1	1	0.2	1	0.8
Wolff-Hirschorn syndrome	0	0.0	8	0.2	5	0.2	1	0.2	1	0.8
Autosomal anomalies in normal infant	0	0.0	140	4.0	112	3.9	13	2.4	1	0.8
Triploidy	1	3.7	34	1.0	15	0.5	0	0.0	0	0.0
Turner syndrome (45,X)	3	11.2	137	3.9	73	2.6	8	1.5	0	0.0
Klinefelter syndrome (47,XXY)	0	0.0	54	1.5	37	1.3	2	0.4	0	0.0
Absent spleen/Asplenia	1	3.7	12	0.3	10	0.4	1	0.2	0	0.0
Accessory spleen	1	3.7	40	1.1	28	1.0	3	0.6	1	0.8
Situs inversus	1	3.7	42	1.2	24	0.8	8	1.5	3	2.4
Conjoined twins	0	0.0	9	0.3	0	0.0	0	0.0	2	1.6
Tuberous sclerosis	1	3.7	14	0.4	12	0.4	1	0.2	1	0.8
Amniotic band syndrome	1	3.7	79	2.3	44	1.6	15	2.8	3	2.4
DiGeorge syndrome	0	0.0	25	0.7	14	0.5	7	1.3	3	2.4
Pierre-Robin sequence	0	0.0	35	1.0	23	0.8	9	1.7	1	0.8
Werdnig-Hoffman syndrome	0	0.0	16	0.5	12	0.4	1	0.2	1	0.8
Wolff-Parkinson-White syndrome	0	0.0	19	0.5	14	0.5	4	0.7	0	0.0
NEOPLASMS	7	26.2	356	10.2	223	7.9	43	8.0	7	5.7
TOTAL BIRTH DEFECT CASES	323	1,207.0	16,449	469.6	11,314	398.6	2,666	492.9	669	541.4

* Rate = (number of birth defects)/(number of live births)x10,000.
\+ Birth weight >2,499 grams.

TABLE 9. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY PRENATAL CARE, HAWAI'I, 1986-2005

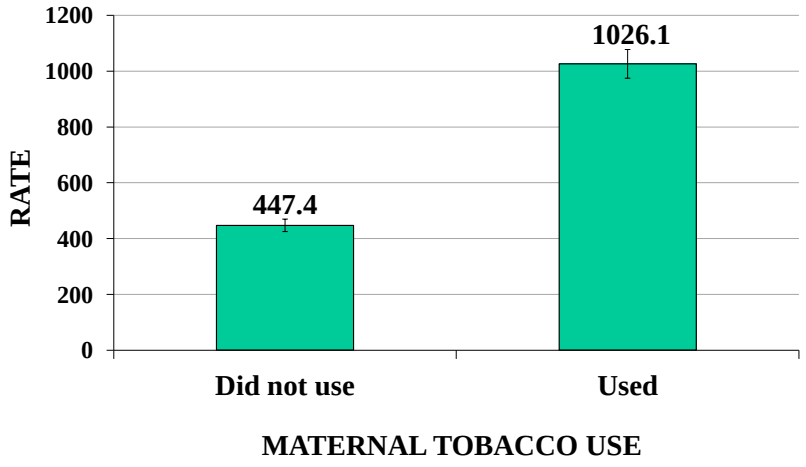
Diagnosis	None N=2,676		Yes-ever N=350,297		Yes-1st Trimester N=283,856		Yes-2nd Trimester N=54,085		Yes-3rd Trimester N=12,356	
	N	Rate*	N	Rate	N	Rate	N	Rate	N	Rate
FETAL ALCOHOL SYNDROME	13	48.6	76	2.2	32	1.1	25	4.6	14	11.3
BIRTH DEFECTS CASES WITH:										
CONGENITAL INFECTION	11	41.1	254	7.3	159	5.6	56	10.4	15	12.1
MATERNAL ALCOHOL USE	151	564.3	1,359	38.8	828	29.2	325	60.1	94	76.1
MATERNAL TOBACCO USE	609	2,275.8	4,355	124.3	2,350	82.8	1,240	229.3	412	333.4
MATERNAL ILLICIT DRUG USE	806	3,012.0	2,347	67.0	989	34.8	842	155.7	333	269.5
Marijuana	169	631.5	832	23.8	416	14.7	282	52.1	82	66.4
Cocaine	147	549.3	436	12.4	191	6.7	156	28.8	54	43.7
Methamphetamine/Amphetamine	651	2,432.7	1,462	41.7	523	18.4	572	105.8	252	203.9

* Rate = (number of birth defects)/(number of live births)x10,000.
+ Birth weight >2,499 grams.

Birth Defect Rates by Maternal Tobacco Use[13]

The birth defect rate for mothers who used tobacco during pregnancy (1,026.1 per 10,000 live births) was over two-fold higher than the rate for mothers who did not use tobacco (447.4) (Figure 10).

FIGURE 10. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY MATERNAL TOBACCO USE, HAWAI'I, 1988-2005



There were also differences among broad categories and specific birth defects for birth defect rates by maternal tobacco use (Table 10).

[13] Birth defect rate by maternal tobacco use: Numerator = number of cases of birth defects whose mother used tobacco (or did not use tobacco) identified through medical record abstraction. Denominator = number of mothers who used tobacco (or did not use tobacco) as listed on the birth certificate (data from DOH OHSM).

TABLE 10. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY MATERNAL TOBACCO USE, HAWAI‘I, 1988-2005*

Diagnosis	Did not use tobacco N=300,636		Used tobacco N=29,665	
	N	Rate**	N	Rate
BRAIN & NERVOUS SYSTEM	1,154	38.4	338	113.9
Neural tube defects	275	9.1	68	22.9
Anencephaly	98	3.3	30	10.1
Spina bifida	128	4.3	26	8.8
Encephalocele	56	1.9	15	5.1
Microcephaly	243	8.1	113	38.1
Holoprosencephaly	36	1.2	10	3.4
Hydrocephaly (w/o spina bifida)	306	10.2	87	29.3
EYE, EAR, FACE, & NECK	1,453	48.3	391	131.8
EYE	543	18.1	152	51.2
Anophthalmia	10	0.3	4	1.3
Microphthalmia	81	2.7	26	8.8
Cataract	34	1.1	10	3.4
EAR	1,014	33.7	275	92.7
Absent auditory canal/ear	40	1.3	5	1.7
Microtia	102	3.4	26	8.8
CARDIAC & CIRCULATORY	5,512	183.3	1,329	448.0
Common truncus	26	0.9	1	0.3
Transposition of the great vessels	126	4.2	32	10.8
Tetralogy of Fallot	113	3.8	25	8.4
Single ventricle	23	0.8	5	1.7
Ventricular septal defect	1,382	46.0	319	107.5
Atrial septal defect	2,121	70.6	510	171.9
Endocardial cushion defect	60	2.0	26	8.8
Pulmonary valve atresia/stenosis	261	8.7	73	24.6
Tricuspid valve atresia/stenosis	43	1.4	16	5.4
Ebstein’s anomaly	13	0.4	3	1.0
Aortic valve atresia/stenosis	35	1.2	14	4.7
Mitral valve atresia/stenosis	27	0.9	10	3.4
Hypoplastic left heart syndrome	46	1.5	12	4.0
Dextrocardia	36	1.2	16	5.4
Patent ductus arteriosus +	2,428	80.8	528	178.0
Coarctation of the aorta	84	2.8	16	5.4
Aortic stenosis	3	0.1	0	0.0
Pulmonary artery atresia/stenosis	560	18.6	140	47.2
Total anomalous pulmonary venous return	32	1.1	5	1.7
Partial anomalous pulmonary venous return	11	0.4	6	2.0
Persistent fetal circulation	165	5.5	36	12.1
RESPIRATORY	714	23.7	194	65.4
Choanal atresia/stenosis	40	1.3	8	2.7
Lung cysts	24	0.8	6	2.0
Hypoplastic lung	162	5.4	41	13.8
OROFACIAL & GASTROINTESTINAL	1,690	56.2	456	153.7
Oral clefts	533	17.7	172	58.0

* Data available beginning 1988.
** Rate = (number of birth defects)/(number of live births)x10,000.
\+ Birth weight >2,499 grams.

TABLE 10. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY MATERNAL TOBACCO USE, HAWAI‘I, 1988-2005*

Diagnosis	Did not use tobacco N=300,636		Used tobacco N=29,665	
	N	Rate**	N	Rate
Cleft palate (isolated)	186	6.2	78	26.3
Cleft lip (+/-cleft palate)	347	11.5	94	31.7
TE fistula/Esophageal atresia	61	2.0	17	5.7
Pyloric stenosis	213	7.1	54	18.2
Atresia/stenosis small intestine	90	3.0	15	5.1
Anal atresia/stenosis	145	4.8	37	12.5
Hirschsprung’s disease	82	2.7	8	2.7
Malrotation	81	2.7	19	6.4
Biliary atresia	31	1.0	7	2.4
GENITAL & URINARY	3,281	109.1	673	226.9
Ovarian cyst	18	0.6	1	0.3
Undescended testicle +	936	31.1	208	70.1
Hypospadias/Epispadias/Chordee	1,065	35.4	158	53.3
Ambiguous genitalia	15	0.5	5	1.7
Absent/hypoplastic kidney	139	4.6	29	9.8
Cystic kidney	129	4.3	29	9.8
Hydronephrosis	426	14.2	77	26.0
Exstrophy bladder	7	0.2	1	0.3
Posterior urethral valves	19	0.6	4	1.3
LIMB & MUSCULOSKELETAL	3,857	128.3	915	308.4
Scoliosis	74	2.5	18	6.1
Hip dislocation	313	10.4	51	17.2
Clubfoot	901	30.0	264	89.0
Polydactyly	506	16.8	121	40.8
Syndactyly	235	7.8	56	18.9
Limb reduction deformity - upper	92	3.1	34	11.5
Limb reduction deformity - lower	41	1.4	12	4.0
Craniosynostosis	141	4.7	26	8.8
Crouzon’s disease	2	0.1	0	0.0
Apert syndrome	10	0.3	1	0.3
Goldenhar syndrome	15	0.5	5	1.7
Spina bifida occulta	8	0.3	3	1.0
Dwarfism	82	2.7	16	5.4
Osteogenesis imperfecta	12	0.4	3	1.0
Diaphragmatic hernia	70	2.3	17	5.7
Abdominal wall defects	246	8.2	64	21.6
Omphalocele	78	2.6	17	5.7
Gastroschisis	96	3.2	32	10.8
Prune-belly syndrome	9	0.3	1	0.3
Poland syndrome	5	0.2	1	0.3
SKIN & INTEGUMENT	1,705	56.7	536	180.7
CHROMOSOME & SYNDROME	1,664	55.3	288	97.1
Down syndrome (trisomy 21)	466	15.5	70	23.6
Patau syndrome (trisomy 13)	62	2.1	6	2.0

* Data available beginning 1988.
** Rate = (number of birth defects)/(number of live births)x10,000.
\+ Birth weight >2,499 grams.

TABLE 10. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY MATERNAL TOBACCO USE, HAWAI'I, 1988-2005*

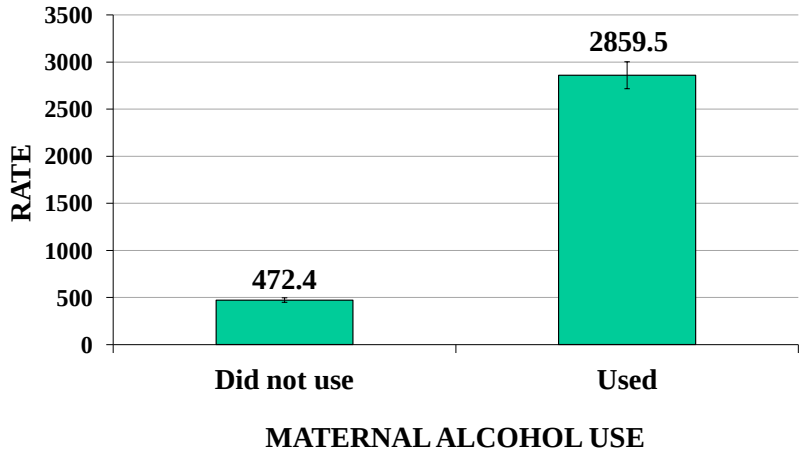
Diagnosis	Did not use tobacco N=300,636		Used tobacco N=29,665	
	N	Rate**	N	Rate
Edwards syndrome (trisomy 18)	140	4.7	20	6.7
Cri du Chat syndrome	7	0.2	1	0.3
Wolff-Hirschorn syndrome	6	0.2	2	0.7
Autosomal anomalies in normal infant	125	4.2	10	3.4
Triploidy	30	1.0	2	0.7
Turner syndrome (45,X)	115	3.8	19	6.4
Klinefelter syndrome (47,XXY)	46	1.5	7	2.4
Absent spleen/Asplenia	7	0.2	6	2.0
Accessory spleen	37	1.2	5	1.7
Situs inversus	34	1.1	9	3.0
Conjoined twins	11	0.4	2	0.7
Tuberous sclerosis	12	0.4	3	1.0
Amniotic band syndrome	55	1.8	25	8.4
DiGeorge syndrome	18	0.6	7	2.4
Pierre-Robin sequence	25	0.8	7	2.4
Werdnig-Hoffman syndrome	13	0.4	3	1.0
Wolff-Parkinson-White syndrome	16	0.5	3	1.0
NEOPLASMS	302	10.0	55	18.5
TOTAL BIRTH DEFECT CASES	13,451	447.4	3,044	1026.1
FETAL ALCOHOL SYNDROME	23	0.8	64	21.6
BIRTH DEFECTS CASES WITH:				
CONGENITAL INFECTION	192	6.4	67	22.6
MATERNAL ALCOHOL USE	3,988	132.7	960	323.6
MATERNAL ILLICIT DRUG USE	823	27.4	2,311	779.0
Marijuana	245	8.1	754	254.2
Cocaine	136	4.5	448	151.0
Methamphetamine/Amphetamine	531	17.7	1,568	528.6

* Data available beginning 1988.
** Rate = (number of birth defects)/(number of live births)x10,000.
\+ Birth weight >2,499 grams.

Birth Defect Rates by Maternal Alcohol Use[14]

The birth defect rate for mothers who used alcohol during pregnancy (2,859.5 per 10,000 live births) was six-fold higher than the rate for mothers who did not use alcohol (472.4) (Figure 11).

FIGURE 11. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY MATERNAL ALCOHOL USE, HAWAI'I, 1988-2005



There were also differences among broad categories and specific birth defects for birth defect rates by maternal alcohol use (Table 11).

[14] Birth defect rate by maternal alcohol use: Numerator = number of cases of birth defects whose mother used alcohol (or did not use alcohol) identified through medical record abstraction. Denominator = number of mothers who used alcohol (or did not use alcohol) as listed on the birth certificate (data from DOH OHSM).

TABLE 11. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY MATERNAL ALCOHOL USE, HAWAI'I, 1988-2005*

Diagnosis	Did not use alcohol N=326,363		Used alcohol N=29,665	
	N	Rate**	N	Rate
BRAIN & NERVOUS SYSTEM	1,369	41.9	120	331.9
Neural tube defects	313	9.6	24	66.4
Anencephaly	114	3.5	9	24.9
Spina bifida	141	4.3	12	33.2
Encephalocele	65	2.0	4	11.1
Microcephaly	311	9.5	46	127.2
Holoprosencephaly	42	1.3	4	11.1
Hydrocephaly (w/o spina bifida)	366	11.2	27	74.7
EYE, EAR, FACE, & NECK	1,670	51.2	182	503.3
EYE	618	18.9	82	226.8
Anophthalmia	11	0.3	3	8.3
Microphthalmia	99	3.0	8	22.1
Cataract	43	1.3	1	2.8
EAR	1,172	35.9	123	340.2
Absent auditory canal/ear	41	1.3	4	11.1
Microtia	117	3.6	10	27.7
CARDIAC & CIRCULATORY	6,420	196.7	410	1,133.8
Common truncus	24	0.7	3	8.3
Transposition of the great vessels	143	4.4	13	36.0
Tetralogy of Fallot	128	3.9	11	30.4
Single ventricle	26	0.8	2	5.5
Ventricular septal defect	1,580	48.4	125	345.7
Atrial septal defect	2,490	76.3	139	384.4
Endocardial cushion defect	78	2.4	8	22.1
Pulmonary valve atresia/stenosis	304	9.3	31	85.7
Tricuspid valve atresia/stenosis	51	1.6	8	22.1
Ebstein's anomaly	15	0.5	1	2.8
Aortic valve atresia/stenosis	42	1.3	6	16.6
Mitral valve atresia/stenosis	34	1.0	3	8.3
Hypoplastic left heart syndrome	54	1.7	4	11.1
Dextrocardia	46	1.4	4	11.1
Patent ductus arteriosus +	2,824	86.5	127	351.2
Coarctation of the aorta	91	2.8	9	24.9
Aortic stenosis	3	0.1	0	0.0
Pulmonary artery atresia/stenosis	662	20.3	39	107.9
Total anomalous pulmonary venous return	36	1.1	1	2.8
Partial anomalous pulmonary venous return	17	0.5	0	0.0
Persistent fetal circulation	194	5.9	6	16.6
RESPIRATORY	824	25.2	81	224.0
Choanal atresia/stenosis	44	1.3	3	8.3
Lung cysts	28	0.9	2	5.5
Hypoplastic lung	191	5.9	11	30.4
OROFACIAL & GASTROINTESTINAL	1,953	59.8	197	544.8
Oral clefts	661	20.3	51	141.0

* Data available beginning 1988.
** Rate=(number of birth defects)/(number of live births)x10,000.
\+ Birth weight >2,499 grams.

TABLE 11. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY MATERNAL ALCOHOL USE, HAWAI'I, 1988-2005*

Diagnosis	Did not use alcohol N=326,363		Used alcohol N=29,665	
	N	Rate**	N	Rate
Cleft palate (isolated)	249	7.6	19	52.5
Cleft lip (+/-cleft palate)	413	12.7	32	88.5
TE fistula/Esophageal atresia	72	2.2	4	11.1
Pyloric stenosis	245	7.5	22	60.8
Atresia/stenosis small intestine	98	3.0	7	19.4
Anal atresia/stenosis	165	5.1	14	38.7
Hirschsprung's disease	85	2.6	3	8.3
Malrotation	98	3.0	2	5.5
Biliary atresia	38	1.2	1	2.8
GENITAL & URINARY	3,734	114.4	212	586.3
Ovarian cyst	18	0.6	1	2.8
Undescended testicle**	1,085	33.2	54	149.3
Hypospadias/Epispadias/Chordee	1,161	35.6	57	157.6
Ambiguous genitalia	19	0.6	1	2.8
Absent/hypoplastic kidney	154	4.7	12	33.2
Cystic kidney	149	4.6	9	24.9
Hydronephrosis	480	14.7	24	66.4
Exstrophy bladder	8	0.2	0	0.0
Posterior urethral valves	23	0.7	0	0.0
LIMB & MUSCULOSKELETAL	4,381	134.2	381	1,053.7
Scoliosis	81	2.5	9	24.9
Hip dislocation	337	10.3	24	66.4
Clubfoot	1,072	32.8	82	226.8
Polydactyly	584	17.9	43	118.9
Syndactyly	260	8.0	30	83.0
Limb reduction deformity - upper	109	3.3	17	47.0
Limb reduction deformity - lower	49	1.5	4	11.1
Craniosynostosis	156	4.8	10	27.7
Crouzon's disease	2	0.1	0	0.0
Apert syndrome	11	0.3	0	0.0
Goldenhar syndrome	19	0.6	2	5.5
Spina bifida occulta	11	0.3	0	0.0
Dwarfism	90	2.8	8	22.1
Osteogenesis imperfecta	15	0.5	0	0.0
Diaphragmatic hernia	81	2.5	5	13.8
Abdominal wall defects	288	8.8	23	63.6
Omphalocele	87	2.7	8	22.1
Gastroschisis	120	3.7	9	24.9
Prune-belly syndrome	10	0.3	0	0.0
Poland syndrome	6	0.2	0	0.0
SKIN & INTEGUMENT	2,021	61.9	221	611.2
CHROMOSOME & SYNDROME	1,797	55.1	152	420.4
Down syndrome (trisomy 21)	492	15.1	43	118.9
Patau syndrome (trisomy 13)	65	2.0	4	11.1

* Data available beginning 1988.
** Rate=(number of birth defects)/(number of live births)x10,000.
\+ Birth weight >2,499 grams.

TABLE 11. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY MATERNAL ALCOHOL USE, HAWAI'I, 1988-2005*

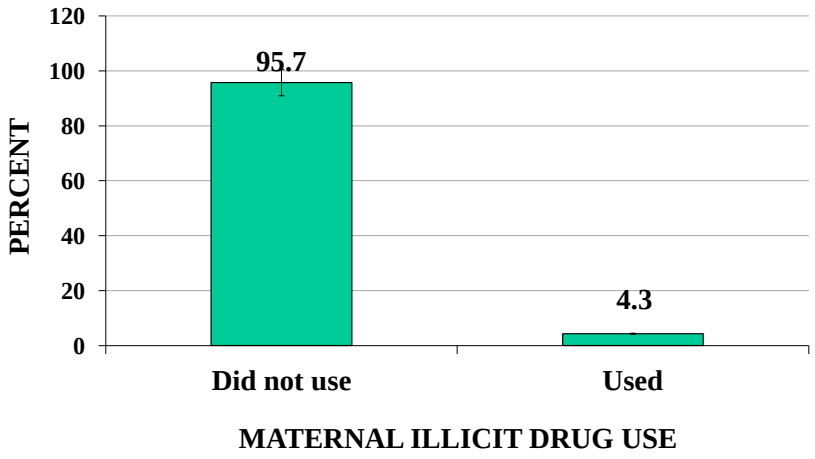
Diagnosis	Did not use alcohol N=326,363		Used alcohol N=29,665	
	N	Rate**	N	Rate
Edwards syndrome (trisomy 18)	152	4.7	8	22.1
Cri du Chat syndrome	6	0.2	2	5.5
Wolff-Hirschorn syndrome	8	0.2	0	0.0
Autosomal anomalies in normal infant	123	3.8	12	33.2
Triploidy	30	0.9	2	5.5
Turner syndrome (45,X)	123	3.8	10	27.7
Klinefelter syndrome (47,XXY)	50	1.5	4	11.1
Absent spleen/Asplenia	13	0.4	0	0.0
Accessory spleen	37	1.1	4	11.1
Situs inversus	42	1.3	1	2.8
Conjoined twins	13	0.4	0	0.0
Tuberous sclerosis	15	0.5	0	0.0
Amniotic band syndrome	65	2.0	15	41.5
DiGeorge syndrome	24	0.7	1	2.8
Pierre-Robin sequence	32	1.0	1	2.8
Werdnig-Hoffman syndrome	15	0.5	1	2.8
Wolff-Parkinson-White syndrome	18	0.6	1	2.8
NEOPLASMS	333	10.2	24	66.4
TOTAL BIRTH DEFECT CASES	15,417	472.4	1,034	2,859.5
FETAL ALCOHOL SYNDROME	6	0.2	83	229.5
BIRTH DEFECTS CASES WITH:				
CONGENITAL INFECTION	241	7.4	16	44.2
MATERNAL TOBACCO USE	561	17.2	960	2,654.9
MATERNAL ILLICIT DRUG USE	2,477	75.9	651	1,800.3
Marijuana	720	22.1	280	774.3
Cocaine	364	11.2	215	594.6
Methamphetamine/Amphetamine	1,766	54.1	333	920.9

* Data available beginning 1988.
** Rate=(number of birth defects)/(number of live births)x10,000.
\+ Birth weight >2,499 grams.

Proportion of Birth Defects by Maternal Illicit Drug Use

For 1986-2005, the proportion of birth defects cases with maternal illicit drug use was 4.3% (Figure 12A). There were also differences among broad categories and specific birth defects by maternal illicit drug use (Table 12).

FIGURE 12A. PROPORTION OF BIRTH DEFECT CASES BY MATERNAL ILLICIT DRUG USE, 1986-2005



For 2000-2005 estimates, the proportion of birth defects for mothers who used illicit drugs during pregnancy (5.8%) was 52% higher than the proportion for mothers who did not use illicit drugs (3.9%) (Figure 12B).

FIGURE 12B. ESTIMATED PROPORTION OF LIVE BIRTHS WITH BIRTH DEFECTS BY MATERNAL ILLICIT DRUG USE, HAWAI'I, 2000-2005

	Maternal illicit drug use	No maternal illicit drug use	Total
Number of live births with birth defects	168	3,964	4,132
Number of live births	2,880 *(est.)* [15]	103,770 *(est.)*[15]	106,650
Proportion of live births with birth defects	5.8% (5.4-6.2) *(est.)*[16]	3.8% (3.4-4.2) *(est.)*[16]	3.9% (3.5-4.3)

[15] Estimate based on data from the Hawai'i State Department of Health, Family Health Services Division, Hawai'i Pregnancy Risk Assessment Monitoring System (PRAMS) Trend Report, 2000-2008. Data showed maternal substance use for 2.7% of women who had a live birth for the years 2004-2008.
[16] Proportion = (live births with birth defects)/(live births)x100.

TABLE 12. PROPORTION OF BIRTH DEFECTS BY MATERNAL ILLICIT DRUG USE, HAWAI‘I, 1986-2005

Diagnosis	Did not use illicit drugs		Used illicit drugs	
	N	% *	N	%
BRAIN & NERVOUS SYSTEM	1,453	94.2	90	5.8
Neural tube defects	353	98.6	5	1.4
Anencephaly	132	99.2	1	0.8
Spina bifida	162	100.0	0	0.0
Encephalocele	69	94.5	4	5.5
Microcephaly	332	89.7	38	10.3
Holoprosencephaly	44	91.7	4	8.3
Hydrocephaly (w/o spina bifida)	384	95.0	20	5.0
EYE, EAR, FACE, & NECK	1,776	93.7	119	6.3
EYE	658	92.5	53	7.5
Anophthalmia	12	85.7	2	14.3
Microphthalmia	97	89.0	12	11.0
Cataract	43	95.6	2	4.4
EAR	1,246	93.5	87	6.5
Absent auditory canal/ear	42	93.3	3	6.7
Microtia	125	96.2	5	3.8
CARDIAC & CIRCULATORY	6,742	95.0	357	5.0
Common truncus	26	96.3	1	3.7
Transposition of the great vessels	154	94.5	9	5.5
Tetralogy of Fallot	132	94.3	8	5.7
Single ventricle	26	92.9	2	7.1
Ventricular septal defect	1,688	95.9	73	4.1
Atrial septal defect	2,613	95.1	135	4.9
Endocardial cushion defect	84	96.6	3	3.4
Pulmonary valve atresia/stenosis	333	95.7	15	4.3
Tricuspid valve atresia/stenosis	56	94.9	3	5.1
Ebstein’s anomaly	18	94.7	1	5.3
Aortic valve atresia/stenosis	49	92.5	4	7.5
Mitral valve atresia/stenosis	36	94.7	2	5.3
Hypoplastic left heart syndrome	56	96.6	2	3.4
Dextrocardia	49	90.7	5	9.3
Patent ductus arteriosus +	2,904	96.1	119	3.9
Coarctation of the aorta	99	94.3	6	5.7
Aortic stenosis	3	100.0	0	0.0
Pulmonary artery atresia/stenosis	689	93.5	48	6.5
Total anomalous pulmonary venous return	37	100.0	0	0.0
Partial anomalous pulmonary venous return	16	94.1	1	5.9
Persistent fetal circulation	206	96.3	8	3.7
RESPIRATORY	882	93.9	57	6.1
Choanal atresia/stenosis	49	98.0	1	2.0
Lung cysts	29	93.5	2	6.5
Hypoplastic lung	203	95.8	9	4.2
OROFACIAL & GASTROINTESTINAL	2,105	94.4	124	5.6
Oral clefts	693	94.0	44	6.0
Cleft palate (isolated)	256	93.1	19	6.9

* Percent = (number of birth defects with or without illicit drug use)/(number of birth defects)x100
+ Birth weight >2,499 grams.

TABLE 12. PROPORTION OF BIRTH DEFECTS BY MATERNAL ILLICIT DRUG USE, HAWAI‘I, 1986-2005

Diagnosis	Did not use illicit drugs		Used illicit drugs	
	N	% *	N	%
Cleft lip (+/-cleft palate)	438	94.6	25	5.4
TE fistula/Esophageal atresia	75	96.2	3	3.8
Pyloric stenosis	276	96.2	11	3.8
Atresia/stenosis small intestine	103	95.4	5	4.6
Anal atresia/stenosis	176	95.7	8	4.3
Hirschsprung’s disease	93	100.0	0	0.0
Malrotation	98	97.0	3	3.0
Biliary atresia	38	95.0	2	5.0
GENITAL & URINARY	3,951	96.5	143	3.5
Ovarian cyst	19	100.0	0	0.0
Undescended testicle +	2,069	98.1	41	1.9
Hypospadias/Epispadias/Chordee	1,241	98.3	22	1.7
Ambiguous genitalia	20	100.0	0	0.0
Absent/hypoplastic kidney	167	96.5	6	3.5
Cystic kidney	160	97.0	5	3.0
Hydronephrosis	502	97.3	14	2.7
Exstrophy bladder	9	100.0	0	0.0
Posterior urethral valves	23	100.0	0	0.0
LIMB & MUSCULOSKELETAL	4,695	95.3	233	4.7
Scoliosis	91	95.8	4	4.2
Hip dislocation	365	97.6	9	2.4
Clubfoot	1,154	95.6	53	4.4
Polydactyly	615	95.6	28	4.4
Syndactyly	274	91.9	24	8.1
Limb reduction deformity - upper	117	90.7	12	9.3
Limb reduction deformity - lower	51	94.4	3	5.6
Craniosynostosis	178	98.3	3	1.7
Crouzon’s disease	2	100.0	0	0.0
Apert syndrome	9	81.8	2	18.2
Goldenhar syndrome	19	90.5	2	9.5
Spina bifida occulta	13	100.0	0	0.0
Dwarfism	98	95.1	5	4.9
Osteogenesis imperfecta	14	93.3	1	6.7
Diaphragmatic hernia	84	95.5	4	4.5
Abdominal wall defects	309	95.7	14	4.3
Omphalocele	98	97.0	3	3.0
Gastroschisis	125	94.7	7	5.3
Prune-belly syndrome	10	100.0	0	0.0
Poland syndrome	6	100.0	0	0.0
SKIN & INTEGUMENT	2,035	88.5	264	11.5
CHROMOSOME & SYNDROME	2,022	96.8	66	3.2
Down syndrome (trisomy 21)	553	97.7	13	2.3
Patau syndrome (trisomy 13)	73	97.3	2	2.7
Edwards syndrome (trisomy 18)	166	98.8	2	1.2
Cri du Chat syndrome	8	100.0	0	0.0

* Percent = (number of birth defects with or without illicit drug use)/(number of birth defects)x100
+ Birth weight >2,499 grams.

TABLE 12. PROPORTION OF BIRTH DEFECTS BY MATERNAL ILLICIT DRUG USE, HAWAI‘I, 1986-2005

Diagnosis	Did not use illicit drugs		Used illicit drugs	
	N	% *	N	%
Wolff-Hirschorn syndrome	7	87.5	1	12.5
Autosomal anomalies in normal infant	137	96.5	5	3.5
Triploidy	46	100.0	0	0.0
Turner syndrome (45,X)	144	96.6	5	3.4
Klinefelter syndrome (47,XXY)	52	94.5	3	5.5
Absent spleen/Asplenia	11	84.6	2	15.4
Accessory spleen	41	97.6	1	2.4
Situs inversus	39	90.7	4	9.3
Conjoined twins	13	100.0	0	0.0
Tuberous sclerosis	14	93.3	1	6.7
Amniotic band syndrome	71	86.6	11	13.4
DiGeorge syndrome	24	96.0	1	4.0
Pierre-Robin sequence	33	91.7	3	8.3
Werdnig-Hoffman syndrome	16	100.0	0	0.0
Wolff-Parkinson-White syndrome	19	95.0	1	5.0
NEOPLASMS	364	96.6	13	3.4
TOTAL BIRTH DEFECT CASES	16,572	95.7	739	4.3
FETAL ALCOHOL SYNDROME	51	56.7	39	43.3
BIRTH DEFECT CASES WITH:				
CONGENITAL INFECTION	254	94.4	15	5.6
MATERNAL ALCOHOL USE	1,328	86.9	200	13.1
MATERNAL TOBACCO USE	4,516	90.2	488	9.8

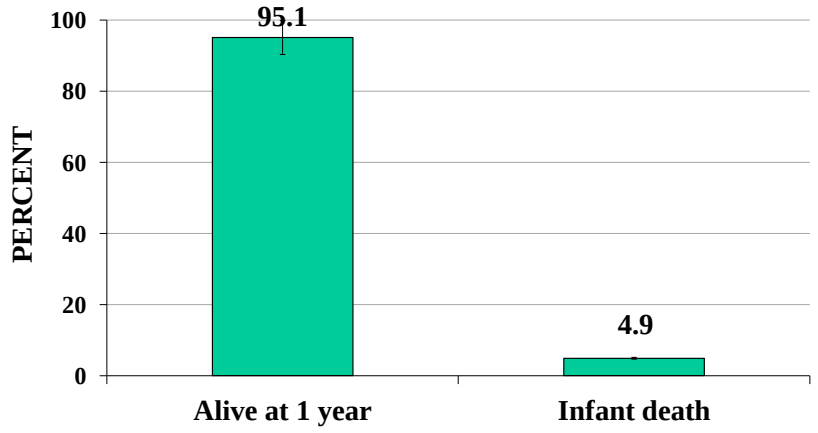
* Percent = (number of birth defects with or without illicit drug use)/(number of birth defects)x100
+ Birth weight >2,499 grams.

Infant Deaths with Birth Defects[17]

Information and further research related to mortality for birth defects may contribute to further developing or improving health, social, and/or family services and support related to infants with birth defects.

Of live births with birth defects, 4.9% were associated with infant death (Figure 13A). There were differences among broad categories and specific birth defects in the proportion associated with infant death (Table 13).

FIGURE 13A. PROPORTION OF LIVE BIRTHS WITH BIRTH DEFECTS ASSOCIATED WITH INFANT DEATH, HAWAI'I, 1986-2005



The number of infant deaths associated with birth defects is 2.1 per 1,000 live births (Figure 13B). Of all infant deaths, 29.6% are associated with birth defects.

FIGURE 13B. INFANT DEATHS ASSOCIATED WITH BIRTH DEFECTS PER 1,000 LIVE BIRTHS, HAWAI'I, 1986-2005

	Number	Number of infant deaths per 1,000 live births*
Live births	370,886	-
Infant deaths – total	2,630	7.1
Infant deaths – with birth defects	774	2.1
Proportion of total infant deaths that have birth defects = 29.6%**		

* Number of infant deaths per 1,000 live births = (infant deaths)/(live births)x1,000
** Proportion = (number of infant deaths with birth defects)/total number of infant deaths)x100

[17] Infant deaths are live births that expired within one year of birth. The presence of a birth defect does not necessarily indicate that the birth defect is the direct cause of death, since there may be other causes of death.

TABLE 13. PROPORTION OF BIRTH DEFECTS ASSOCIATED WITH INFANT DEATH, HAWAI'I, 1986-2005

Diagnosis	Live births	Alive at 1 year		Infant deaths*	
	N	N	% **	N	%
BRAIN & NERVOUS SYSTEM	1,224	1,034	84.5	190	15.5
Neural tube defects	173	127	73.4	46	26.6
Anencephaly	23	0	0.0	23	100.0
Spina bifida	119	107	89.9	12	10.1
Encephalocele	35	20	57.1	15	42.9
Microcephaly	362	310	85.6	52	14.4
Holoprosencephaly	32	17	53.1	15	46.9
Hydrocephaly (w/o spina bifida)	318	258	81.1	60	18.9
EYE, EAR, FACE, & NECK	1,773	1,546	87.2	227	12.8
EYE	688	594	86.3	94	13.7
Anophthalmia	8	4	50.0	4	50.0
Microphthalmia	106	67	63.2	39	36.8
Cataract	45	42	93.3	3	6.7
EAR	1,260	1,072	85.1	188	14.9
Absent auditory canal/ear	45	41	91.1	4	8.9
Microtia	126	116	92.1	10	7.9
CARDIAC & CIRCULATORY	6,908	6,344	91.8	564	8.2
Common truncus	22	12	54.5	10	45.5
Transposition of the great vessels	152	119	78.3	33	21.7
Tetralogy of Fallot	133	115	86.5	18	13.5
Single ventricle	25	16	64.0	9	36.0
Ventricular septal defect	1,710	1,581	92.5	129	7.5
Atrial septal defect	2,695	2,406	89.3	289	10.7
Endocardial cushion defect	76	56	73.7	20	26.3
Pulmonary valve atresia/stenosis	340	306	90.0	34	10.0
Tricuspid valve atresia/stenosis	57	48	84.2	9	15.8
Ebstein's anomaly	17	15	88.2	2	11.8
Aortic valve atresia/stenosis	52	34	65.4	18	34.6
Mitral valve atresia/stenosis	38	16	42.1	22	57.9
Hypoplastic left heart syndrome	47	20	42.6	27	57.4
Dextrocardia	51	38	74.5	13	25.5
Patent ductus arteriosus +	3,015	2,854	94.7	161	5.3
Coarctation of the aorta	103	89	86.4	14	13.6
Aortic stenosis	3	2	66.7	1	33.3
Pulmonary artery atresia/stenosis	731	712	97.4	19	2.6
Total anomalous pulmonary venous return	37	28	75.7	9	24.3
Partial anomalous pulmonary venous return	16	13	81.3	3	18.8
Persistent fetal circulation	214	172	80.4	42	19.6
RESPIRATORY	879	669	76.1	210	23.9
Choanal atresia/stenosis	50	46	92.0	4	8.0
Lung cysts	29	25	86.2	4	13.8
Hypoplastic lung	184	46	25.0	138	75.0
OROFACIAL & GASTROINTESTINAL	2,112	1,937	91.7	175	8.3
Oral clefts	673	620	92.1	53	7.9
Cleft palate (isolated)	258	238	92.2	20	7.8

* Expired within 1 year of birth.
** Percent = (number alive or with infant death)/(number of live births)x100.
\+ Birth weight >2,499 grams.

TABLE 13. PROPORTION OF BIRTH DEFECTS ASSOCIATED WITH INFANT DEATH, HAWAI‘I, 1986-2005

Diagnosis	Live births	Alive at 1 year		Infant deaths*	
	N	N	% **	N	%
Cleft lip (+/-cleft palate)	415	382	92.0	33	8.0
TE fistula/Esophageal atresia	74	59	79.7	15	20.3
Pyloric stenosis	285	283	99.3	2	0.7
Atresia/stenosis small intestine	101	88	87.1	13	12.9
Anal atresia/stenosis	172	147	85.5	25	14.5
Hirschsprung’s disease	93	84	90.3	9	9.7
Malrotation	93	79	84.9	14	15.1
Biliary atresia	38	35	92.1	3	7.9
GENITAL & URINARY	3,969	3,749	94.5	220	5.5
Ovarian cyst	19	19	100.0	0	0.0
Undescended testicle +	1,168	1,148	98.3	20	1.7
Hypospadias/Epispadias/Chordee	1,250	1,222	97.8	28	2.2
Ambiguous genitalia	14	12	85.7	2	14.3
Absent/hypoplastic kidney	140	93	66.4	47	33.6
Cystic kidney	145	113	77.9	32	22.1
Hydronephrosis	493	466	94.5	27	5.5
Exstrophy bladder	8	8	100.0	0	0.0
Posterior urethral valves	17	14	82.4	3	17.6
LIMB & MUSCULOSKELETAL	4,666	4,368	93.6	298	6.4
Scoliosis	74	54	73.0	20	27.0
Hip dislocation	372	359	96.5	13	3.5
Clubfoot	1,161	1,098	94.6	63	5.4
Polydactyly	620	600	96.8	20	3.2
Syndactyly	283	252	89.0	31	11.0
Limb reduction deformity - upper	112	85	75.9	27	24.1
Limb reduction deformity - lower	47	34	72.3	13	27.7
Craniosynostosis	181	170	93.9	11	6.1
Crouzon’s disease	2	2	100.0	0	0.0
Apert syndrome	11	8	72.7	3	27.3
Goldenhar syndrome	21	19	90.5	2	9.5
Spina bifida occulta	11	9	81.8	2	18.2
Dwarfism	88	71	80.7	17	19.3
Osteogenesis imperfecta	14	8	57.1	6	42.9
Diaphragmatic hernia	76	43	56.6	33	43.4
Abdominal wall defects	250	217	86.8	33	13.2
Omphalocele	58	41	70.7	17	29.3
Gastroschisis	114	107	93.9	7	6.1
Prune-belly syndrome	9	5	55.6	4	44.4
Poland syndrome	6	6	100.0	0	0.0
SKIN & INTEGUMENT	2,275	2,166	95.2	109	4.8
CHROMOSOME & SYNDROME	1,238	1,052	85.0	186	15.0
Down syndrome (trisomy 21)	328	306	93.3	22	6.7
Patau syndrome (trisomy 13)	28	6	21.4	22	78.6
Edwards syndrome (trisomy 18)	54	18	33.3	36	66.7
Cri du Chat syndrome	5	5	100.0	0	0.0

* Expired within 1 year of birth.
** Percent = (number alive or with infant death)/(number of live births)x100.
\+ Birth weight >2,499 grams.

TABLE 13. PROPORTION OF BIRTH DEFECTS ASSOCIATED WITH INFANT DEATH, HAWAI'I, 1986-2005

Diagnosis	Live births	Alive at 1 year		Infant deaths*	
	N	N	% **	N	%
Wolff-Hirschorn syndrome	7	7	100.0	0	0.0
Autosomal anomalies in normal infant	130	127	97.7	3	2.3
Triploidy	0	0	-	0	-
Turner syndrome (45,X)	37	36	97.3	1	2.7
Klinefelter syndrome (47,XXY)	35	35	100.0	0	0.0
Absent spleen/Asplenia	8	7	87.5	1	12.5
Accessory spleen	29	3	10.3	26	89.7
Situs inversus	40	35	87.5	5	12.5
Conjoined twins	2	0	0.0	2	100.0
Tuberous sclerosis	15	15	100.0	0	0.0
Amniotic band syndrome	62	44	71.0	18	29.0
DiGeorge syndrome	25	19	76.0	6	24.0
Pierre-Robin sequence	36	35	97.2	1	2.8
Werdnig-Hoffman syndrome	13	7	53.8	6	46.2
Wolff-Parkinson-White syndrome	20	19	95.0	1	5.0
NEOPLASMS	233	214	91.8	19	8.2
TOTAL BIRTH DEFECT CASES	15,815	15,041	95.1	774	4.9
FETAL ALCOHOL SYNDROME	87	84	96.6	3	3.4
BIRTH DEFECTS CASES WITH:					
CONGENITAL INFECTION	250	229	91.6	21	8.4
MATERNAL ALCOHOL USE	1,399	1,330	95.1	69	4.9
MATERNAL TOBACCO USE	4,776	4,607	96.5	169	3.5
MATERNAL ILLICIT DRUG USE	3,081	3,018	98.0	63	2.0
Marijuana	973	948	97.4	25	2.6
Cocaine	565	556	98.4	9	1.6
Methamphetamine/Amphetamine	2,074	2,045	98.6	29	1.4

* Expired within 1 year of birth.
** Percent = (number alive or with infant death)/(number of live births)x100.
\+ Birth weight >2,499 grams.

Comparison of Birth Defect Rates for Hawai‘i and U.S.

A comparison of birth defect rates for Hawai‘i (2000-2005) and the U.S. (2004-2006) indicates that Hawai‘i rates for anencephaly, encephalocele, and transposition of great arteries are higher than U.S. rates (Figure 14). It also indicates that Hawai‘i rates for other birth defects are similar to or lower than U.S. rates.

Figure 14. Comparison of Birth Defect Rates for Hawai‘i (2000-2005) and U.S. (2004-2006)

	Hawai‘i (2000-2005)[18]			**US (2004-2006)**[19]	
Birth Defect	Average Annual Number of Cases	Hawai‘i Prevalence per 10,000 Live Births	Confidence Interval	Estimated Annual Number of Cases	Est. National Prevalence per 10,000 Live Births
					Adjusted for maternal race/ethnicity
Central nervous system defects					
Anencephaly	6	3.0	2.7 – 3.3	859	2.06
Spina bifida without anencephaly	5	2.8	2.5 – 3.1	1,460	3.50
Encephalocele	2	1.3	1.1 – 1.5	341	0.82
Eye defects					
Anophthalmia/microphthalmia	4	1.9	1.6 – 2.2	780	1.87
Cardiovascular defects					
Common truncus	1	0.3	0.2 – 0.4	301	0.72
Transposition of great arteries	8	4.5	4.1 – 4.8	1,252	3.00
Tetralogy of Fallot	5	2.8	2.5 – 3.1	1,657	3.97
Atrioventricular septal defect (endocardial cushion defect)	4	2.0	1.7 – 2.2	1,966	4.71
Hypoplastic left heart syndrome	2	0.9	0.7 – 1.1	960	2.30
Orofacial defects					
Cleft palate without cleft lip	11	6.1	5.7 – 6.5	2,651	6.35
Cleft lip with and without cleft palate	17	9.0	8.4 – 9.5	4,437	10.63
Gastrointestinal defects					
Esophageal atresia/ tracheoesophageal fistula	3	1.4	1.2 – 1.6	905	2.17
Rectal and large intestinal atresia/stenosis	5	2.8	2.5 – 3.1	1,952	4.68
Musculoskeletal defects					
Reduction deformity, upper limbs	5	2.5	2.2 – 2.8	1,454	3.49
Reduction deformity, lower limbs	3	1.3	1.1 – 1.5	701	1.68
Gastroschisis	7	3.8	3.4 – 4.1	1,871	4.49
Omphalocele	3	1.8	1.6 – 2.0	775	1.86
Diaphragmatic hernia	2	1.2	1.0 – 1.4	1,088	2.61
					Adjusted for maternal age
Chromosomal anomalies					
Trisomy 13	3	1.4	1.2 – 1.6	528	1.26
Trisomy 21 (Down syndrome)	22	11.8	11.2 – 12.4	6,037	14.47
Trisomy 18	5	2.7	2.4 – 3.0	1,109	2.66

[18] Data source: HBDP birth defects surveillance data.
[19] Data source: Parker SE, Mai CT, Canfield MA, et al. Updated National Birth Prevalence Estimates for Selected Birth Defects in the United States, 2004-2006. *Birth Defects Research (Part A): Clin. Mol.Teratol.* 2010;88:1008-1016.

APPENDIX

APPENDIX A
BRITISH PEDIATRIC ASSOCIATION CODES
FOR SPECIFIC DIAGNOSES

DIAGNOSIS	CODES
BRAIN & NERVOUS SYSTEM	740.000-742.999
NEURAL TUBE DEFECTS	740.000-742.099
Anencephaly	740.000-740.199
Spina bifida	741.000-741.999
Encephalocele	742.000-742.099
MICROCEPHALY	742.100-742.199
HOLOPROSENCEPHALY	742.260-742.269
HYDROCEPHALY (W/O SPINA BIFIDA)	742.300-742.399
EYE, EAR, FACE & NECK	743.000-744.999
EYE	743.000-743.999
Anophthalmia	743.000-743.099
Microphthalmia	743.100-743.199
Cataract	743.320-743.326
EAR	744.000-744.499
Absent auditory canal/ear	744.000-744.019
Microtia	744.210-744.219
CARDIAC & CIRCULATORY	745.000-747.999
COMMON TRUNCUS	745.000-745.099
TRANSPOSITION GREAT VESSELS	745.100-745.199
TETRALOGY OF FALLOT	745.200-745.299
SINGLE VENTRICLE	745.300-745.399
VENTRICULAR SEPTAL DEFECT	745.400-745.499
ATRIAL SEPTAL DEFECT	745.500-745.599
ENDOCARDIAL CUSHION DEFECT	745.600-745.699
PULMONARY VALVE ATRESIA/STENOSIS	746.000-746.019 746.995
TRICUSPID VALVE ATRESIA/STENOSIS	746.100-746.104
EBSTEIN'S ANOMALY	746.200-746.299
AORTIC VALVE ATRESIA/STENOSIS	746.300-746.399 746.480-746.489
MITRAL VALVE ATRESIA/STENOSIS	746.500-746.599
HYPOPLASTIC LEFT HEART SYNDROME	746.700-746.799
DEXTROCARDIA	746.800-746.809
PATENT DUCTUS ARTERIOSUS†	747.000-747.099
COARCTATION AORTA	747.100-747.199
AORTA STENOSIS	747.200-747.209
PULMONARY ARTERY ATRESIA/STENOSIS	747.300-747.329
TOTAL ANOMALOUS PULMONARY VENOUS RETURN	747.420-747.429
PARTIAL ANOMALOUS PULMONARY VENOUS RETURN	747.430-747.439
PERSISTENT FETAL CIRCULATION	747.900-747.999

† Birth weight >2499 gm

RESPIRATORY	748.000-748.999
CHOANAL ATRESIA/STENOSIS	748.000-748.099
LUNG CYSTS	748.400-748.499
HYPOPLASTIC LUNG	748.510-748.519
OROFACIAL & GASTROINTESTINAL	749.000-751.999
ORAL CLEFTS	749.000-749.299
Cleft palate (isolated)	749.000-749.099
Cleft lip (±cleft palate)	749.100-749.299
TRACHEOESOPHAGEAL FISTULA/ESOPHAGEAL ATRESIA	750.300-750.399
PYLORIC STENOSIS	750.510-750.519
ATRESIA/STENOSIS SMALL INTESTINE	751.100-751.199
ANAL ATRESIA/STENOSIS	751.200-751.299
HIRSCHSPRUNG'S DISEASE	751.300-751.399
MALROTATION	751.400-751.499
BILIARY ATRESIA	751.650-751.659
GENITAL & URINARY	752.000-753.999
OVARIAN CYST	752.080-752.089
UNDESCENDED TESTICLE†	752.500-752.599
HYPOSPADIAS/EPISPADIAS/CHORDEE	752.600-752.699
AMBIGUOUS GENITALIA	752.790-752.799
ABSENT/HYPOPLASTIC KIDNEY	753.000-753.099
CYSTIC KIDNEY	753.100-753.199
HYDRONEPHROSIS	753.200-753.209
EXSTROPHY BLADDER	753.500-753.599
POSTERIOR URETHRAL VALVES	753.600-753.609
LIMB & MUSCULOSKELETAL	754.000-756.999
SCOLIOSIS	754.200-754.299
HIP DISLOCATION	754.300-754.309
CLUBFOOT	754.500-754.799
POLYDACTYLY	755.500-755.099
SYNDACTYLY	755.100-755.199
LIMB REDUCTION DEFORMITY-UPPER	755.200-755.299
LIMB REDUCTION DEFORMITY-LOWER	755.300-755.399
CRANIOSYNOSTOSIS	756.000-756.039
CROUZON'S DISEASE	756.040
APERT SYNDROME	756.050-756.059
GOLDENHAR SYNDROME	756.060-756.069
SPINA BIFIDA OCCULTA	756.100-756.109
DWARFISM	756.400-756.499
OSTEOGENESIS IMPERFECTA	756.500-756.509
DIAPHRAGMATIC HERNIA	756.610-756.619
ABDOMINAL WALL DEFECT	756.700-756.799
Omphalocele	756.700-756.709
Gastroschisis	756.710-756.719
Prune-belly syndrome	756.720-756.729
POLAND SYNDROME	756.800-756.809

† Birth weight >2499 gm

SKIN & INTEGUMENT	757.000-757.999
CHROMOSOME & OTHER SYNDROME	758.000-759.999
DOWN SYNDROME (TRISOMY 21)	758.000-758.099
PATAU SYNDROME (TRISOMY 13)	758.100-758.199
EDWARDS SYNDROME (TRISOMY 18)	758.200-758.299
CRI DU CHAT SYNDROME	758.310-758.319
WOLFF-HIRSCHORN SYNDROME	758.320-758.329
AUTOSOMAL ANOMALIES IN NORMAL INFANT	758.400-758.499
TRIPLOIDY	758.586
TURNER SYNDROME (45, X)	758.600-758.699
KLINEFELTER SYNDROME (47, XXY)	758.700-758.799
ABSENT SPLEEN/ASPLENIA	759.000-759.009
ACCESSORY SPLEEN	759.040-759.049
SITUS INVERSUS	759.300-759.399
CONJOINED TWINS	759.400-759.499
TUBEROUS SCLEROSIS	759.500-759.599
AMNIOTIC BAND SYNDROME	658.800-658.899
DIGEORGE SYNDROME	279.110-279.119
PEIRRE-ROBIN SEQUENCE	524.080-524.089
WERDNIG-HOFFMAN SYNDROME	335.000-335.999
WOLFF-PARKINSON-WHITE SYNDROME	426.705
NEOPLASMS	155.000-214.999
	228.100-239.999
CONGENITAL INFECTION	042.000-042.999
	090.000-090.999
	771.000-771.299
	774.480-774.499
MATERNAL SUBSTANCE ABUSE	760.700-760.799
FETAL ALCOHOL SYNDROME	760.710-760.719
ILLICIT DRUGS	760.720-760.799
Cocaine	760.720-760.799
Marijuana	760.720-760.799
Methamphetamine/Amphetamine	760.720-760.799

APPENDIX B
PARTICIPATING HEALTH CARE FACILITIES

NAME	ISLAND
Castle Medical Center	Oahu
Fetal Diagnostic Institute of the Pacific	Oahu
Hana Medical Center	Maui
Hilo Medical Center	Hawai‘i
Hale Ho‘ola Hamakua Hospital (formerly Honoka‘a)*	Hawai‘i
Kahuku Hospital	Oahu
Kaiser-Permanente Medical Care	Oahu
Kapiolani Medical Center (KMC)	Oahu
KMC Fetal Diagnostic Center	Oahu
KMC Genetics Section**	Oahu
KMC Molecular and Clinical Cytogenetics Laboratory**	Oahu
Kau Hospital	Hawai‘i
Kauai Veterans Memorial Hosp	Kauai
Kohala Hospital	Hawai‘i
Kona Community Hospital	Hawai‘i
Kula Hospital*	Maui
Lanai Community Hospital	Lanai
Maui Memmorial Hospital	Maui
Medical Genetics Services**	Oahu
Mid-Pacific Genetics**	Oahu
Molokai General Hospital	Molokai
North Hawai‘i Community Hospital	Hawai‘i
Pali Momi Medical Center	Oahu
Queens Medical Center (QMC)	Oahu
QMC Comprehensive Genetics Center	Oahu
QMC Genetics Laboratory	Oahu
Shriner’s Hospital for Children	Oahu
St. Francis Medical Center	Oahu
St. Francis Medical Center-West*	Oahu
Straub Clinic and Hospital	Oahu
Tripler Army Medical Center	Oahu
Wahiawa General Hospital	Oahu
Waianae Coast Comprehensive Health Center	Oahu
G.N. Wilcox Memorial Hospital	Kauai

*No longer has deliveries
**No longer in operation

Mahalo!

HBDP appreciates the assistance from the DOH Office of Health Status Monitoring (OHSM) in providing data for this surveillance report. OHSM provided data on live births, fetal deaths, infant deaths, births with alcohol use, births with tobacco use, and other data used in the calculations of birth defect rates and proportions.

HAWAIʻI BIRTH DEFECTS PROGRAM
Children with Special Health Needs Branch
Family Health Services Division
Hawaiʻi State Department of Health
741 Sunset Avenue
Honolulu, Hawaiʻi 96816

Website:
http://hawaii.gov/health/family-child-health/genetics/hbdhome.html

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For help with a problem, please contact the Hawaiʻi State Department of Health at 1250 Punchbowl Street, Room 216, Honolulu, HI 96813 or (808) 586-4122 within 180 days. You may also contact our departmental Affirmative Action Officer at P.O. Box 3378, Honolulu, HI 96801-3378 or (808) 586-4616 (voice/tty).