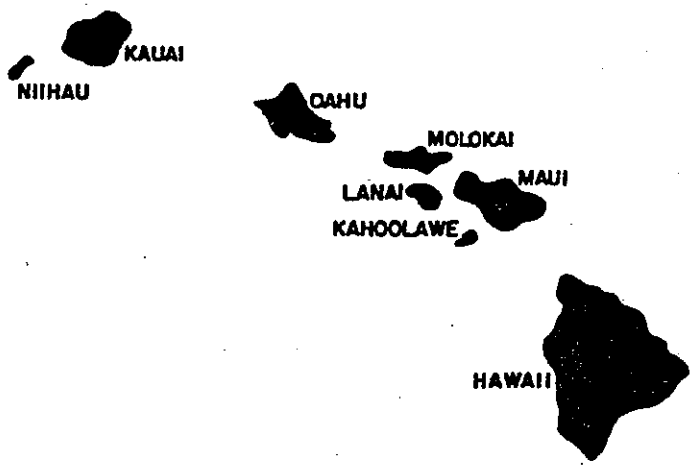
HAWAII BIRTH DEFECTS PROGRAM

1986-2003 STATEWIDE DATA



SURVEILLANCE REPORT NUMBER 12

ON BIRTH DEFECTS IN HAWAII

JANUARY 1, 1986 - DECEMBER 31, 2003

For: December 2004

Governor of Hawaii

Linda Lingle

Director of Health

Chiyome Leinaala Fukino, MD

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LINDA LINGLE
GOVERNOR OF HAWAII

CHIYOME L. FUKINO, M.D.
DIRECTOR OF HEALTH

STATE OF HAWAII
DEPARTMENT OF HEALTH
FAMILY HEALTH SERVICES DIVISION
CHILDREN WITH SPECIAL HEALTH NEEDS BRANCH
HAWAII BIRTH DEFECTS PROGRAM
76 NORTH KING STREET, #208 • HONOLULU, HAWAII 96817-5157
TEL:808-587-4120 • FAX: 808-587-4130 • E-MAIL: hbdp@crch.hawaii.edu

To Our Readers,

The Hawaii Birth Defects Program is very pleased to present its twelfth surveillance report on birth defects diagnosed in the State of Hawaii for the eighteen years between January 1, 1986, and December 31, 2003.

We hope that this publication is informative and helpful to you, and we welcome any comments or suggestions. Requests for more specific data can be made by writing the Registry at the address shown above.

Sincerely,

Ruth D. Merz, M.S.
Administrator
Hawaii Birth Defects Program

Mathias B. Forrester, B.S.
Data Coordination Consultant
Hawaii Birth Defects Program

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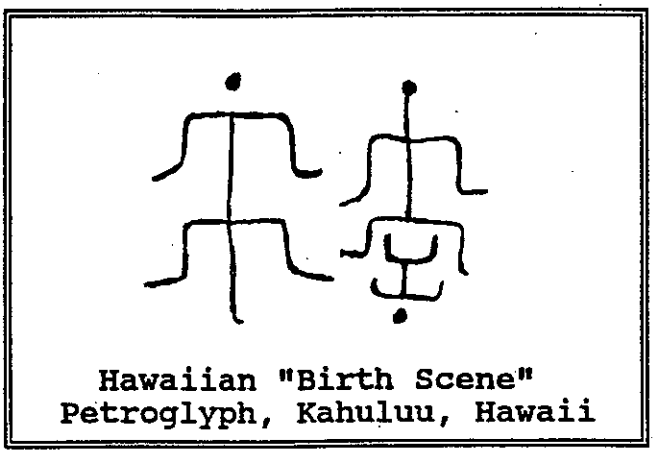

Hawaiian "Birth Scene"
Petroglyph, Kahuluu, Hawaii

INTRODUCTION

Purpose and Goals of the HBDP:

The Hawaii Birth Defects Program (HBDP) is a population-based, multiple ascertainment source, active surveillance system (registry) for birth defects and other adverse pregnancy outcomes covering the entire state of Hawaii.

A birth defect is defined as any structural, functional, or biochemical abnormality in development that originates before birth and is detectable at birth or shortly thereafter. These defects may be genetically linked or caused by environmental hazards or adverse life style effects, but most often are of unknown origin. Birth defects are the number one cause of fetal death and infant mortality in Hawaii each year. About one in five (20%) of all pregnancies result in spontaneous miscarriage and over 1,000 (5.2%) of all babies born each year in Hawaii are born with some recognizable abnormality. According to the Centers for Disease Control and Prevention (CDC), approximately 30% of all admissions to pediatric hospitals are caused by problems directly associated with birth defects. These statistics represent a significant impact for Hawaii's families, with birth defects touching many people either directly or indirectly.

The purpose of the HBDP is to be a reliable, valid, and timely information source for ascertaining the number of infants and young children (up to one year of age) with specific disabilities, the number of pregnancies resulting in adverse reproductive outcomes, and for providing relevant and timely information and data to community agencies, hospitals, and state departments for policy, planning, and decision making purposes.

With eighteen years of retrospectively and prospectively secured data and in-depth statistical analyses, the HBDP has examined elevated adverse reproductive outcomes, possible geographic and other clusters, and birth defect trends and changes over time. The Program also provides information for case-control studies and for developing public awareness education programs about birth defects and their causes. And lastly, the HBDP acts as a resource for the planning and development of appropriate statewide and community level services and preventive strategies.

History of the HBDP:

The 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 Hawaii. Since its inception seventeen years ago, the HBDP has been a population based, multiple ascertainment (34 facilities/organizations), active

surveillance system serving all of the Hawaiian islands.

Statutory Authority:

From 1988 until 2002, according to an Executive Chamber Ruling by Governor John Waihee dated June 16, 1989, "Hawaii Revised Statutes, sections 321-31 and 338-2 in conjunction, provide the statutory authority for the Department of Health to administer a birth defects monitoring program and to collect statistical data on birth defects." Additional legislative authority was also provided through Hawaii Revised Statutes 324-1 and 324-2 (1990 Amendments). Then, during the 2002 Hawaii Legislative Session, Act 252 (SB 2763, SD2, HD2, CD1 - Relating to Birth Defects) was enacted by the legislature, and signed by the Governor on 7/1/02, which amended Chapters 321- and 324- and Section 572-5 of the Hawaii Revised Statues authorizing the Department of Health to establish the statewide Hawaii Birth Defects Program and registry, increase the marriage license fee from $50 to $60, of which $10 was to be allocated to the Hawaii birth defects special fund, and authorize an expenditure ceiling for the special fund.

HBDP Advisory Committee, Staffing and Funding:

A 20 member HBDP Advisory Committee (composed of professionals from the medical, university, community, public and private sector) offer scientific guidance and input into the Program. HBDP staff consist of four full time field and office based employees as well as a part-time data coordination consultant. Funding is secured through the Birth Defects Special Fund, contracts, and grants from both the public and private sectors.

Administration of the HBDP:

The HBDP operates under the Hawaii State Department of Health (DOH), Family Health Services Division, Children With Special Health Needs Branch.

Patient Eligibility:

In order for an infant/fetus to be included in the HBDP, it must meet the following criteria:

- 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 Centers for Disease Control and Prevention (CDC), as well as other adverse neonatal conditions such as congenital infections, neonatal tumors, fetal alcohol syndrome (FAS), and antenatal use of illicit substances/drugs by the mother.

- The end of the pregnancy must occur in the state of Hawaii, but conception and/or any part of the pregnancy may occur outside of the state.

- The infant or fetus must have been noted to have had a problem during the antenatal period or within one year after delivery. However, definitive diagnoses may be made after the one-year cut-off period. Therefore, statistics for 2003 births may not be complete until after January 2004. However, with few exceptions, most defects indicate their presence prior to or around the time of delivery, and 2003 statistics in this publication are considered to be complete for most defects. The single exception to this policy is fetal alcohol syndrome (FAS). Due to the public health interest in FAS and the difficulty in its diagnosis within the neonatal period, all cases of FAS are included in the Program, no matter what age the diagnosis is made.

All infants and fetuses are included regardless of pregnancy outcome (live birth, fetal demise, or therapeutic termination).

Data Sources and Collection Methods:

Potential cases are identified through examination of discharge lists of certain ICD-9 diagnosis codes and other reports provided on a regular basis by all hospitals in Hawaii where births typically occur, as well as at certain tertiary care sites, diagnostic clinics and laboratories, and other facilities in the state (see Appendix A). Participating health care facilities also include all cytogenetics laboratories, genetic counseling centers, and maternal-fetal centers in the state. Due to its nature as an island archipelago in the middle of the ocean, residents of Hawaii are less likely to travel to other states to deliver their babies, than residents of the rest of the country.

Program staff review both the infant's and mother's medical records to determine if the case meets registry criteria. If it does, the staff complete a data abstraction form in the field on laptop computers. This form permits collection of over 125 pieces of demographic and health related information on each infant/fetus and his/her biological parents, including maternal and paternal race/ethnicity, the number and type of previous pregnancies affected by birth defects, and maternal use of alcohol, tobacco, and recreational drugs during pregnancy (either by self-report, physician note, or positive toxicology screen). If the record indicates that the infant or fetus or his/her mother were seen at another health care facility, the records are reviewed at all other facilities to ensure completeness of the data. In the office, the data collection staff download the data to the office main computer. Office and/or medical personnel then review 100% of the data for completeness and accuracy of coding before transferring the information to the main data base.

Approximately five percent of cases are re-abstracted as part of the Program's quality control/assurance measures.

Denominators for calculation of basic defect rates are provided by the Department of Health's Office of Health Status Monitoring. Since its inception, the HBDP has entered over 19,198 cases (44,025 abstracts) into its registry. In sixteen years, the Program has completed eighteen years of data collection for the period 1986 through 2003, and is in the process of collecting data for the year 2004.

Birth Defect Categories:

The HBDP functions as a registry for all infants with moderate to severe birth defects, as well as for other adverse neonatal conditions such as prenatal maternal substance abuse, neoplasms, and congenital infections. The HBDP codes this diagnostic information using a modified version of the British Pediatric Association system, which is similar to the International Classification of Diseases (ICD), but which allows for greater specificity. This coding system is hierarchical in nature with a greater number of digits corresponding to a more precise diagnosis. The HBDP collects the most precise level possible for diagnoses that fall primarily within the ICD 740.00-759.99 range, plus other congenital anomaly codes specified in the CDC Birth Defects and Genetic Diseases Branch June 1989 publication. The ICD codes reviewed by the HBDP are presented in Appendix B.

Quality Control/Assurance:

To ensure accuracy, strict quality control/assurance procedures have been established as follows: 5% re-abstraction of all medical record charts (by HBDP field staff) and 100% review by office and/or medical personnel (certified dysmorphologist) of all abstracts for completeness and accuracy.

Confidentiality:

Comprehensive control procedures have been established to protect the privacy of families, infants, physicians, and hospitals. Any identifying information is kept strictly confidential and the HBDP is in compliance with all hospital Institutional Review Board (IRB) rules as well as the Health Insurance Portability and Accountability Act of 1996 (HIPAA) regulations passed to date. All HBDP related personnel have taken a written oath of confidentiality, while all data are reported to the community in an aggregate de-identified format only.

Collaboration and Information Dissemination:

The HBDP has assisted new and established USA and international birth defects registries with information, provided data to the CDC and the National Birth Defects Prevention Network for their special studies and publications, and carried out its own research, culminating in published reports and articles in professional peer-reviewed journals. At the local level, the HBDP Administrator serves on the March of Dimes Chapter of the Pacific Community Program Planning as well as the Needs Assessment Committees, the Fetal Alcohol Syndrome Coalition, the Child Death Review Council and its Data Committee, the Hawaii Perinatal Consortium Data Committee, and the Prenatal Nutrition and Folic Acid/Neural Tube Defects Council. Nationally, the HBDP Administrator has served on the National Birth Defects Prevention Network (NBDPN) Executive Committee, chaired the Surveillance Guidelines and Standards Committee charged with developing a Registry User Manual, and is currently serving on the NBDPN Membership and Elections Committee as well as the Data and Research Committee. At NBDPN conferences, the HBDP Administrator has given presentations, done poster sessions, facilitated round table discussions, chaired committee planning meetings, served as MC for the new members orientation and chaired the annual elections process. From a national perspective, site visits by the CDC have been successfully completed by the HBDP several times.

Since 1988, the Program has been approached by members of the community, students in medicine, nursing and public health, physicians, health care institutions and hospitals, the March of Dimes Foundation, and the Hawaii Department of Health for information. The HBDP has provided data on a wide variety of topics including rates of all birth defects, neural tube defects, trisomy 21 and other major chromosome anomalies, heart defects, congenital infections, fetal alcohol syndrome, and maternal alcohol, tobacco, and illicit drug use. The HBDP Administrator gives presentations to both the lay and professional communities, and prepares articles for publications, including 45 appearing in professional peer-review journals. Since 1995, the HBDP has participated in one international (WHO), 7 national (CDC/NBDPN), and 5 local (DOH cluster investigation) special data studies. All of these outreach activities have enhanced the dissemination of information on the many facets of birth defects in Hawaii.

What Makes the HBDP Unique:

The HBDP is able to identify a much higher percentage of infants and fetuses with birth defects and other problems than the only other system currently in use in the state (DOH birth and death certificates). This is consistent with national studies which have observed that active ascertainment statewide birth defect surveillance programs to be a more complete source of birth defect information than the passive birth certificate system.

The state of Hawaii has an ethnic mix unique to the United States, with the majority of the population being non-white. Recently, concern has been raised on the applicability of the results of research conducted on whites to non-whites. Review of HBDP data allow validation of this prior research. Moreover, Hawaii's Program subdivides the Asian/Pacific Islander ethnic group into subgroups such as Japanese, Chinese, Korean, Vietnamese, Cambodian, Filipino, Guamanian/Chammoro, native Hawaiian, and Samoan, among others. Thus the possibility exists to compare particular Asian/Pacific Islander groups with one another and other ethnic groups. The HBDP is adaptable and thus able to collaborate with other organizations on research projects of interest to both.

National Recognition and Rating of the HBDP:

In 1999, the Pew Environmental Health Commission at the Johns Hopkins School of Public Health in their publication entitled *"Healthy From The Start"* gave the Hawaii Birth Defects Program an "A" grade and designated it as one of the top eight programs in the country, and one of only four nationwide to meet all eight rating criteria. Likewise in 2002, the Trust for America's Health in their publication entitled *"Birth Defects Tracking and Prevention: Too Many States Are Not Making the Grade"* also gave the HBDP an "A" grade and rated it as one of the top eight programs in the nation. Of the eight states receiving an "A" grade, four of those other states were cited as needing improvement in at least one of the four major "Categories" and more specific twenty rating "Criteria", while Hawaii was not.

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Data and Information For This Report Were Collected, Analyzed and Prepared For Publication By The Following HBDP Staff Members:

Ruth D. Merz, M.S
Administrator

Mathias B. Forrester, B.S.
Data Coordination Consultant (Part-Time)

Edward R. Diaz, B.S.
Database Associate

A. Michelle Weaver, C.C.H.W.
Data Collection Specialist

Amy M. Yamamoto, C.C.H.W.
Data Collection Specialist

Contacting the HBDP:

The HBDP may be contacted at its office site as follows:

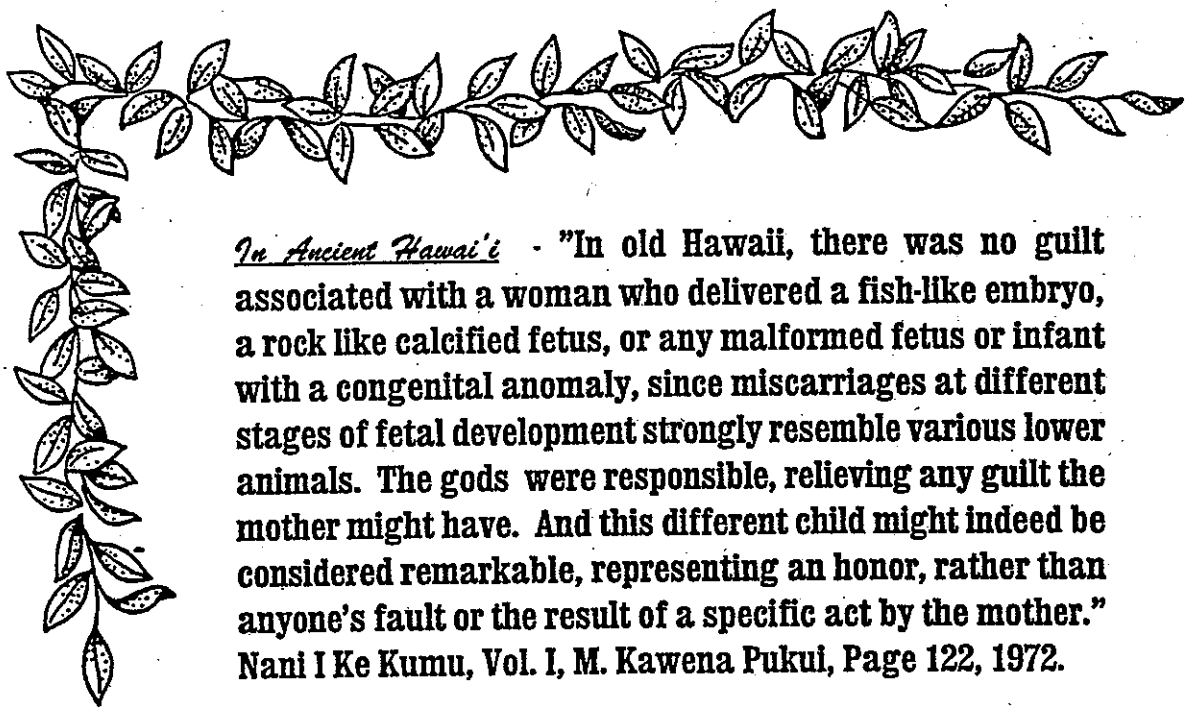
Administrator:	Ruth D. Merz
Office Location/ Mailing Address:	76 North King Street, #208 Honolulu, Hawaii 96817-5157
Telephone:	808-587-4120 (Hawaiian Standard Time)
Fax:	808-587-4130
E-Mail:	hbdp@crch.hawaii.edu

HAWAII BIRTH DEFECTS PROGRAM STAFF

Edward R. Diaz, Amy M.M. Yamamoto, Ruth D. Merz, and A. Michelle Weaver

PREVALENCE OF BIRTH DEFECTS IN HAWAII

The HBDP data base contains 16,573 cases of infants/fetuses delivered in Hawaii between 1986 and 2002 fitting its adverse reproductive outcome diagnostic criteria. Of these cases, 13,965 had birth defects, while the remaining cases involved adverse health conditions not considered CDC designated birth defects, (i.e., neoplasms, congenital infections, and maternal substance abuse during pregnancy), but which the HBDP Program includes in its data base to meet state interests and local concerns. The rates for each year for total adverse reproductive outcomes as well as cases with birth defects are presented in Table 1.



In Ancient Hawai'i - **"In old Hawaii, there was no guilt associated with a woman who delivered a fish-like embryo, a rock like calcified fetus, or any malformed fetus or infant with a congenital anomaly, since miscarriages at different stages of fetal development strongly resemble various lower animals. The gods were responsible, relieving any guilt the mother might have. And this different child might indeed be considered remarkable, representing an honor, rather than anyone's fault or the result of a specific act by the mother." Nani I Ke Kumu, Vol. I, M. Kawena Pukui, Page 122, 1972.**

TABLE 1. PROPORTION OF ALL BIRTHS WITH ADVERSE BIRTH CONDITIONS AND BIRTH DEFECTS, HAWAII, 1986-2003

YEAR	ADVERSE REPRODUCTIVE OUTCOME CASES		BIRTH DEFECTS	
	N	Percent	N	Percent
1986	881	4.4	825	4.2
1987	1092	5.4	1027	5.1
1988	1056	5.1	979	4.8
1989	1094	5.2	943	4.5
1990	1103	4.9	983	4.4
1991	1004	4.6	903	4.1
1992	974	4.5	868	4.0
1993	1078	5.1	932	4.4
1994	1199	5.8	1007	4.8
1995	1197	6.1	1024	5.2
1996	1302	6.7	1135	5.9
1997	1124	6.2	925	5.1
1998	1020	5.5	848	4.5
1999	833	4.6	694	3.8
2000	897	4.9	734	4.0
2001	932	5.2	735	4.1
2002	875	4.8	699	3.8
2003	894	4.7	720	3.8
Total	18,555	5.2	15,981	4.5

FIGURE 1. PROPORTION OF ALL BIRTHS WITH ADVERSE BIRTH CONDITIONS, HAWAII, 1986-2003

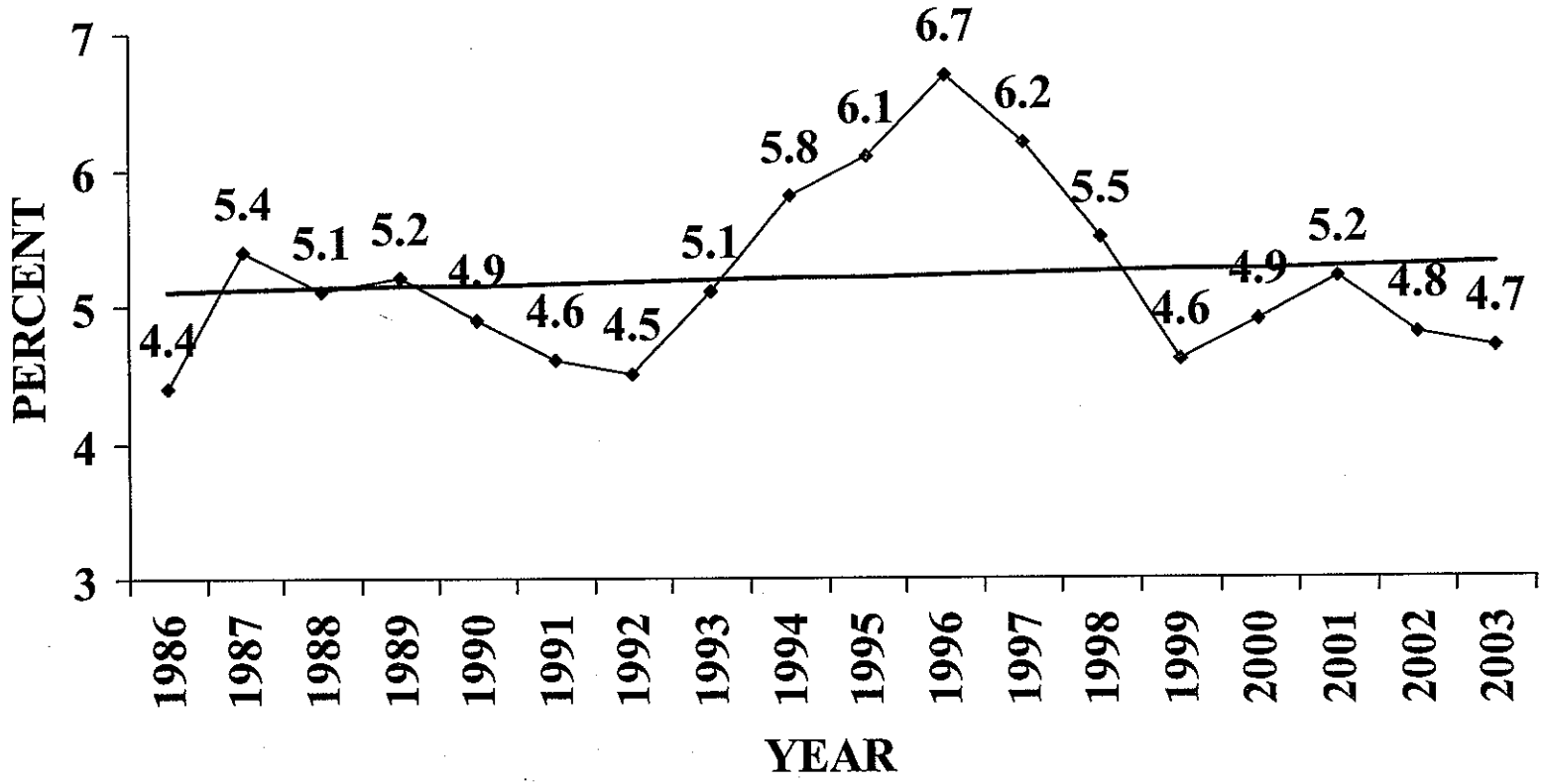
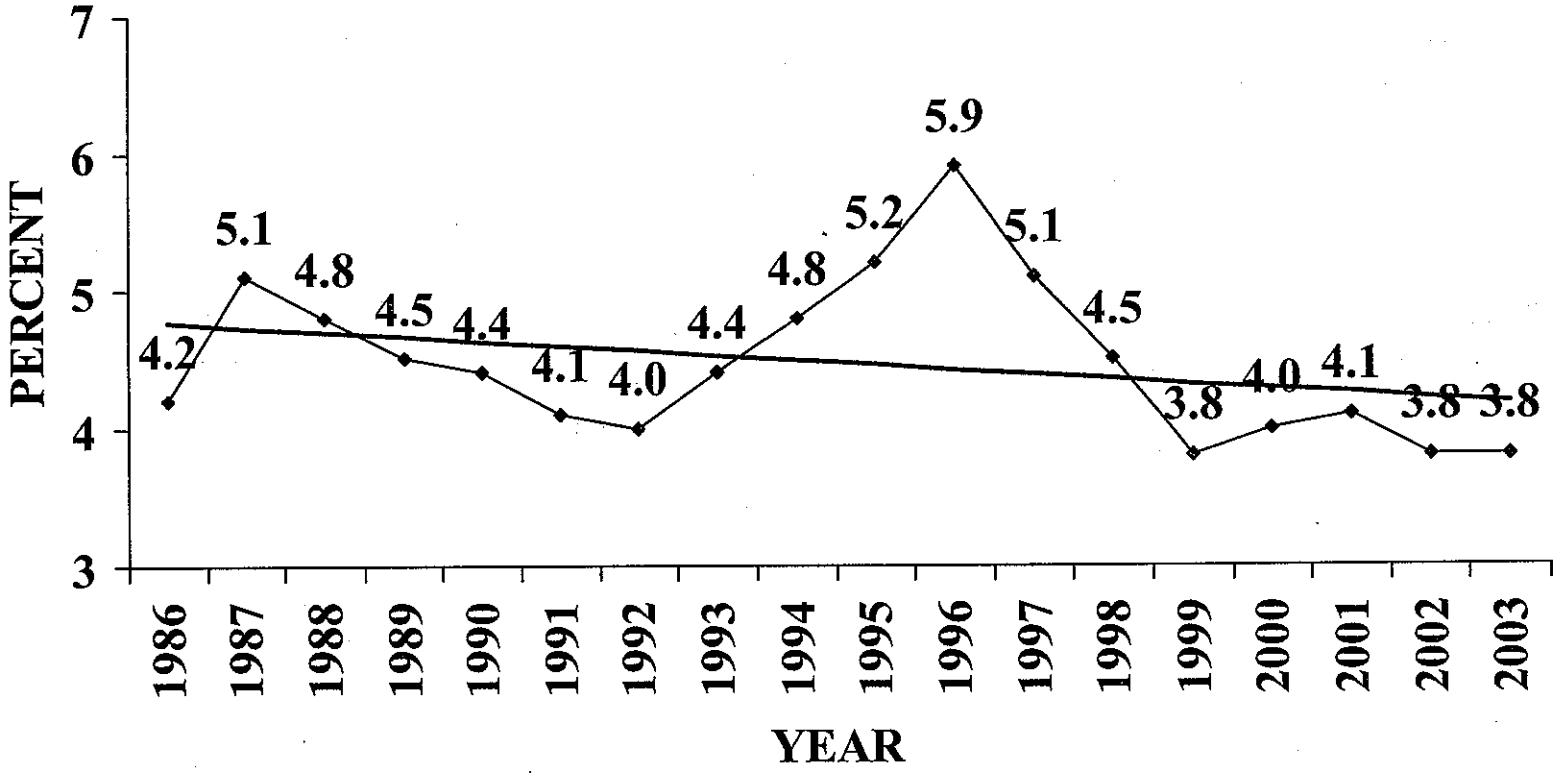


FIGURE 2. PROPORTION OF ALL BIRTHS WITH BIRTH DEFECTS, HAWAII, 1986-2003

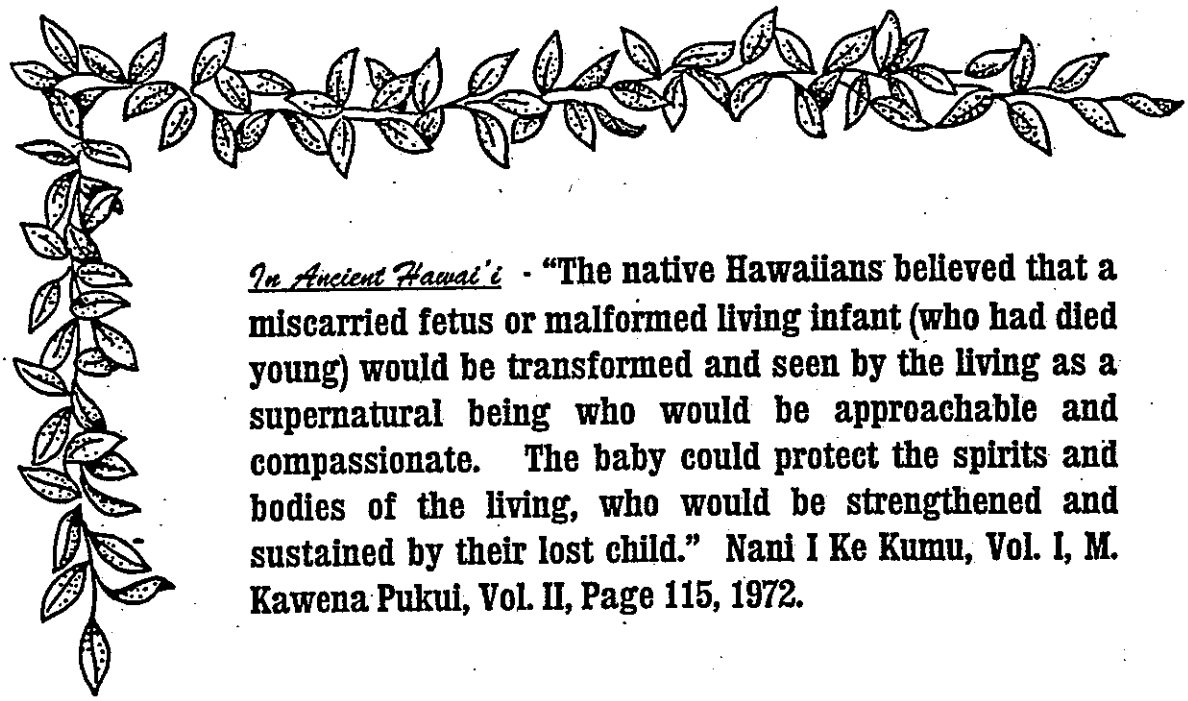


PREVALENCE OF SPECIFIC BIRTH DEFECTS IN HAWAII (COMPARISON WITH OTHER STATES)

Table 2 contains the number and prevalence of selected birth defects in Hawaii from 1986 to 2002 and compares the rates with those from birth defects registries in other states.

Comparisons between the various states should be made with caution. Some of the registries practice active surveillance methods while others are passive. Registries may differ in their classifications of certain defects. (The 6-digit British Pediatric Association system diagnostic codes used for specific defects in the HBDP are provided in Appendix C.) Also, the data from the various registries cover different time periods, and the prevalence of certain birth defects has been found to vary over time. Moreover, registries may differ in what type of pregnancy outcomes are eligible for inclusion in their programs. Some states do not keep track of terminations due to fetal anomalies, and those states which include fetal demises may have cut-off ages or weights.

Please also note that for all data in this report, when numbers are small, statistical measures may not be as useful as when larger numbers are utilized. Slight changes in small numbers can lead to large changes in the measure, making them less stable and useful. Caution should be maintained when making decisions or recommendations based on small numbers.



In Ancient Hawai'i - **"The native Hawaiians believed that a miscarried fetus or malformed living infant (who had died young) would be transformed and seen by the living as a supernatural being who would be approachable and compassionate. The baby could protect the spirits and bodies of the living, who would be strengthened and sustained by their lost child." Nani I Ke Kumu, Vol. I, M. Kawena Pukui, Vol. II, Page 115, 1972.**

TABLE 2. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS, HAWAII, 1986-2003 (COMPARED TO OTHER STATES)*

DIAGNOSIS	Hawaii 1986-2003		AR 97-01	CA 97-01	GA 97-01	IA 97-01	NJ 97-01	NY 97-01	NC 97-00	WI 97-01
	356,460		184,731	283,066	235,616	187,312	571,846	1,281,686	452,582	340,351
	N	Rate	Rate	Rate	Rate	Rate	Rate	Rate	Rate	Rate
BRAIN & NERVOUS SYSTEM	**1424**	**39.9**	-	-	-	-	-	-	-	-
Neural tube defects	333	9.3	-	-	-	-	-	-	-	-
Anencephaly	120	3.4	4.06	3.36	3.40	3.10	0.40	0.40	1.97	1.43
Spina bifida	154	4.3	5.31	4.10	3.57	6.03	1.85	2.27	4.95	2.87
Encephalocele	68	1.9	1.41	0.71	1.36	1.23	0.40	0.66	1.35	0.61
Microcephaly	346	9.7	7.31	-	7.94	9.61	5.14	4.63	6.65	1.70
Holoprosencephaly	42	1.2	-	-	-	-	-	-	-	-
Hydrocephaly (w/o spina bifida)	385	10.8	12.34	2.76	8.15	10.89	3.99	7.21	11.00	3.64
EYE, EAR, FACE, & NECK	**1798**	**50.4**	-	-	-	-	-	-	-	-
EYE	686	19.2	-	-	-	-	-	-	-	-
Anophthalmia	13	0.4	-	-	-	-	-	-	-	-
Microphthalmia	104	2.9	-	-	-	-	-	-	-	-
Cataract	44	1.2	3.09	-	2.16	3.10	1.29	1.20	1.83	1.02
EAR	1264	35.5	-	-	-	-	-	-	-	-
Absent auditory canal/ear	37	1.0	-	-	-	-	-	-	-	-
Microtia	121	3.4	-	-	-	-	-	-	-	-
CARDIAC & CIRCULATORY	**6460**	**181.2**	-	-	-	-	-	-	-	-
Common truncus	24	0.7	0.70	0.39	0.76	1.49	0.63	0.61	1.04	0.29
Transposition of the great vessels	147	4.1	4.60	4.10	5.35	5.55	3.71	3.62	4.73	1.61
Tetralogy of Fallot	131	3.7	3.95	3.67	3.73	3.79	3.90	4.25	5.37	1.61
Single ventricle	28	0.8	-	-	-	-	-	-	-	-
Ventricular septal defect	1627	45.6	38.87	15.65	39.85	44.47	51.41	34.57	41.30	20.27
Atrial septal defect	2371	66.5	-	-	-	-	-	-	-	-
Endocardial cushion defect	81	2.3	4.49	4.13	4.41	5.50	2.82	2.57	4.13	1.43
Pulmonary valve atresia/stenosis	331	9.3	10.23	1.10	7.00	11.85	14.32	8.19	12.53	5.37
Tricuspid valve atresia/stenosis	56	1.6	1.19	4.10	1.66	1.17	0.84	0.86	1.15	0.38
Ebstein's anomaly	18	0.5	0.87	0.28	0.42	0.85	0.42	0.55	0.51	0.26

TABLE 2. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS, HAWAII, 1986-2003 (COMPARED TO OTHER STATES)*

DIAGNOSIS	Hawaii 1986-2003		AR 97-01	CA 97-01	GA 97-01	IA 97-01	NJ 97-01	NY 97-01	NC 97-00	WI 97-01
	356,460		184,731	283,066	235,616	187,312	571,846	1,281,686	452,582	340,351
	N	Rate	Rate	Rate	Rate	Rate	Rate	Rate	Rate	Rate
Aortic valve atresia/stenosis	51	1.4	3.74	1.52	2.08	4.11	1.24	1.37	2.03	0.52
Mitral valve atresia/stenosis	37	1.0	-	-	-	-	-	-	-	-
Hypoplastic left heart syndrome	53	1.5	3.14	2.23	3.06	3.58	2.13	1.84	3.03	1.29
Dextrocardia	54	1.5	-	-	-	-	-	-	-	-
Patent ductus arteriosus**	2703	75.8	-	-	-	-	-	-	-	-
Coarctation of the aorta	97	2.7	5.09	3.89	5.48	7.42	3.15	3.84	5.70	1.20
Aorta stenosis	2	0.1	-	-	-	-	-	-	-	-
Pulmonary artery atresia/stenosis	642	18.0	-	-	-	-	-	-	-	-
Total anomalous pulmonary venous return	32	0.9	-	-	-	-	-	-	-	-
Partial anomalous pulmonary venous	17	0.5	-	-	-	-	-	-	-	-
Persistent fetal circulation	201	5.6	-	-	-	-	-	-	-	-
RESPIRATORY	**909**	**25.5**	-	-	-	-	-	-	-	-
Choanal atresia/stenosis	50	1.4	-	-	-	-	-	-	-	-
Lung cysts	28	0.8	-	-	-	-	-	-	-	-
Hypoplastic lung	209	5.9	-	-	-	-	-	-	-	-
OROFACIAL & GASTROINTESTINAL	**2082**	**58.4**	-	-	-	-	-	-	-	-
Oral clefts	680	19.1	-	-	-	-	-	-	-	-
Cleft palate (isolated)	249	7.0	7.04	4.45	7.09	7.69	5.42	5.84	6.34	5.96
Cleft lip (±cleft palate)	432	12.1	11.42	9.93	8.40	12.55	7.64	7.12	9.59	8.22
TE fistula/Esophageal atresia	71	2.0	2.54	1.20	2.38	2.94	2.33	2.21	3.40	1.61
Pyloric stenosis	272	7.6	12.78	-	13.41	31.50	17.84	17.07	20.77	0.11
Atresia/stenosis small intestine	96	2.7	-	-	-	-	-	-	-	-
Anal atresia/stenosis	171	4.8	5.68	2.72	4.03	5.02	3.46	3.86	5.46	3.58
Hirschsprung's disease	84	2.4	1.14	-	2.33	2.40	1.80	1.97	3.25	0.52
Malrotation	97	2.7	-	-	-	-	-	-	-	-
Biliary atresia	36	1.0	0.70	0.74	0.76	0.80	0.30	0.94	2.14	0.05
GENITAL & URINARY	**3791**	**106.4**	-	-	-	-	-	-	-	-

TABLE 2. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS, HAWAII, 1986-2003 (COMPARED TO OTHER STATES)*

DIAGNOSIS	Hawaii 1986-2003		AR 97-01	CA 97-01	GA 97-01	IA 97-01	NJ 97-01	NY 97-01	NC 97-00	WI 97-01
	356,460		184,731	283,066	235,616	187,312	571,846	1,281,686	452,582	340,351
	N	Rate	Rate	Rate	Rate	Rate	Rate	Rate	Rate	Rate
Ovarian cyst	18	0.5	-	-	-	-	-	-	-	-
Undescended testicle**	1138	31.9	-	-	-	-	-	-	-	-
Hypospadias/Epispadias/Chordee	1141	32.0	-	-	-	-	-	-	-	-
Ambiguous genitalia	15	0.4	-	-	-	-	-	-	-	-
Absent/hypoplastic kidney	167	4.7	4.82	0.57	4.37	5.71	3.29	2.90	4.77	2.73
Cystic kidney	154	4.3	-	-	-	-	-	-	-	-
Hydronephrosis	436	12.2	-	-	-	-	-	-	-	-
Exstrophy bladder	9	0.3	0.27	-	0.13	0.59	0.24	0.23	0.60	0.52
Posterior urethral valves	22	0.6	-	-	-	-	-	-	-	-
LIMB & MUSCULOSKELETAL	**4638**	**130.1**	-	-	-	-	-	-	-	-
Scoliosis	90	2.5	-	-	-	-	-	-	-	-
Hip dislocation	360	10.1	-	-	-	-	-	-	-	-
Clubfoot	1127	31.6	-	-	-	-	-	-	-	-
Polydactyly	602	16.9	-	-	-	-	-	-	-	-
Syndactyly	285	8.0	-	-	-	-	-	-	-	-
Limb reduction deformity - upper	119	3.3	3.74	3.50	4.03	5.61	3.04	1.69	3.03	2.52
Limb reduction deformity - lower	51	1.4	1.73	1.59	1.74	3.15	1.63	1.17	1.92	1.41
Craniosynostosis	175	4.9	-	-	-	-	-	-	-	-
Crouzon's disease	2	0.1	-	-	-	-	-	-	-	-
Apert syndrome	11	0.3	-	-	-	-	-	-	-	-
Goldenhar syndrome	19	0.5	-	-	-	-	-	-	-	-
Spina bifida occulta	12	0.3	-	-	-	-	-	-	-	-
Dwarfism	96	2.7	-	-	-	-	-	-	-	-
Osteogenesis imperfecta	15	0.4	-	-	-	-	-	-	-	-
Diaphragmatic hernia	85	2.4	4.17	2.33	2.29	2.24	2.08	1.19	3.40	2.20
Abdominal wall defects	301	8.4	-	-	-	-	-	-	-	-
Omphalocele	95	2.7	2.54	0.95	2.50	2.83	1.29	1.07	1.67	-

TABLE 2. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS, HAWAII, 1986-2003 (COMPARED TO OTHER STATES)*

DIAGNOSIS	Hawaii 1986-2003		AR 97-01	CA 97-01	GA 97-01	IA 97-01	NJ 97-01	NY 97-01	NC 97-00	WI 97-01
	356,460		184,731	283,066	235,616	187,312	571,846	1,281,686	452,582	340,351
	N	Rate	Rate	Rate	Rate	Rate	Rate	Rate	Rate	Rate
Gastroschisis	118	3.3	4.98	3.89	2.16	4.16	1.82	1.15	4.19	4.14
Prune-belly syndrome	9	0.3	-	-	-	-	-	-	-	-
Poland syndrome	5	0.1	-	-	-	-	-	-	-	
SKIN & INTEGUMENT	**2158**	**60.5**	-	-	-	-	-	-	-	-
CHROMOSOME & SYNDROME	**1925**	**54.0**	-	-	-	-	-	-	-	-
Down syndrome (trisomy 21)	520	14.6	11.91	12.51	13.03	14.47	11.23	10.63	12.42	9.90
Patau syndrome (trisomy 13)	67	1.9	0.92	0.92	1.44	1.49	0.65	0.79	1.86	0.67
Edwards syndrome (trisomy 18)	161	4.5	1.62	1.77	2.55	2.40	0.96	0.15	2.32	1.02
Cri du Chat syndrome	8	0.2	-	-	-	-	-	-	-	-
Wolff-Hirschorn syndrome	8	0.2	-	-	-	-	-	-	-	-
Autosomal anomalies in normal infant	137	3.8	-	-	-	-	-	-	-	-
Triploidy	44	1.2	-	-	-	-	-	-	-	-
Turner syndrome (45,X)	140	3.9	-	-	-	-	-	-	-	-
Klinefelter syndrome (47,XXY)	51	1.4	-	-	-	-	-	-	-	-
Absent spleen/Asplenia	11	0.3	-	-	-	-	-	-	-	-
Accessory spleen	41	1.2	-	-	-	-	-	-	-	-
Situs inversus	37	1.0	-	-	-	-	-	-	-	-
Conjoined twins	12	0.3	-	-	-	-	-	-	-	-
Tuberous sclerosis	13	0.4	-	-	-	-	-	-	-	-
Amniotic band syndrome	77	2.2	2.22	1.80	1.49	1.60	0.49	0.18	-	-
DiGeorge syndrome	21	0.6	-	-	-	-	-	-	-	-
Pierre-Robin sequence	31	0.9	-	-	-	-	-	-	-	-
Werdnig-Hoffman syndrome	14	0.4	-	-	-	-	-	-	-	-
Wolff-Parkinson-White syndrome	19	0.5	-	-	-	-	-	-	-	-
NEOPLASMS	**358**	**10.0**	-	-	-	-	-	-	-	-
CONGENITAL INFECTION	**267**	**7.5**	-	-	-	-	-	-	-	-
MATERNAL SUBSTANCE ABUSE	**2871**	**80.5**	-	-	-	-	-	-	-	-

TABLE 2. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS, HAWAII, 1986-2003 (COMPARED TO OTHER STATES)*

DIAGNOSIS	Hawaii 1986-2003		AR 97-01	CA 97-01	GA 97-01	IA 97-01	NJ 97-01	NY 97-01	NC 97-00	WI 97-01
	356,460		184,731	283,066	235,616	187,312	571,846	1,281,686	452,582	340,351
	N	Rate	Rate	Rate	Rate	Rate	Rate	Rate	Rate	Rate
Fetal alcohol syndrome	89	2.5	1.68	0.60	1.36	0.48	0.35	1.12	-	1.93
Illicit drugs	2820	79.1	-	-	-	-	-	-	-	-
Marijuana	924	25.9	-	-	-	-	-	-	-	-
Cocaine	576	16.2	-	-	-	-	-	-	-	-
Methamphetamine/Amphetamine	1826	51.2	-	-	-	-	-	-	-	-

*Other state rates from National Birth Defects Prevention Network. Birth defects surveillance data from selected states, 1997-2001. Birth Defects Res Part A Clin Mol Teratol 2004;70:677-771.

**birth weight >2499 grams.

TABLE 3. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY YEAR, HAWAII, 1986-2003

DIAGNOSIS	1986		1987		1988		1989		1990		1991		1992		1993	
	19,819		20,097		20,541		20,954		22,288		21,765		21,504		21,073	
	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
BRAIN & NERVOUS SYSTEM	**74**	**37.3**	**74**	**36.8**	**73**	**35.5**	**86**	**41.0**	**85**	**38.1**	**83**	**38.1**	**87**	**40.5**	**100**	**47.5**
Neural tube defects	16	8.1	18	9.0	13	6.3	21	10.0	18	8.1	11	5.1	30	14.0	26	12.3
Anencephaly	5	2.5	11	5.5	5	2.4	5	2.4	8	3.6	5	2.3	10	4.7	8	3.8
Spina bifida	9	4.5	5	2.5	6	2.9	10	4.8	5	2.2	4	1.8	17	7.9	15	7.1
Encephalocele	3	1.5	2	1.0	3	1.5	7	3.3	5	2.2	3	1.4	3	1.4	3	1.4
Microcephaly	27	13.6	24	11.9	25	12.2	28	13.4	17	7.6	27	12.4	17	7.9	26	12.3
Holoprosencephaly	1	0.5	2	1.0	1	0.5	2	1.0	3	1.3	2	0.9	1	0.5	1	0.5
Hydrocephaly (w/o spina bifida)	26	13.1	21	10.4	20	9.7	20	9.5	30	13.5	26	11.9	21	9.8	17	8.1
EYE, EAR, FACE, & NECK	**105**	**53.0**	**108**	**53.7**	**109**	**53.1**	**116**	**55.4**	**110**	**49.4**	**117**	**53.8**	**96**	**44.6**	**89**	**42.2**
EYE	34	17.2	37	18.4	49	23.9	49	23.4	48	21.5	48	22.1	43	20.0	34	16.1
Anophthalmia	0	0.0	1	0.5	0	0.0	1	0.5	1	0.4	0	0.0	0	0.0	2	0.9
Microphthalmia	2	1.0	7	3.5	8	3.9	10	4.8	9	4.0	8	3.7	4	1.9	6	2.8
Cataract	0	0.0	3	1.5	5	2.4	3	1.4	1	0.4	2	0.9	4	1.9	1	0.5
EAR	85	42.9	74	36.8	81	39.4	87	41.5	82	36.8	84	38.6	66	30.7	63	29.9
Absent auditory canal/ear	1	0.5	0	0.0	4	1.9	1	0.5	4	1.8	4	1.8	0	0.0	2	0.9
Microtia	8	4.0	10	5.0	4	1.9	7	3.3	8	3.6	5	2.3	8	3.7	1	0.5
CARDIAC & CIRCULATORY	**280**	**141.3**	**428**	**213.0**	**343**	**167.0**	**327**	**156.1**	**314**	**140.9**	**319**	**146.6**	**244**	**113.5**	**303**	**143.8**
Common truncus	1	0.5	2	1.0	4	1.9	1	0.5	2	0.9	0	0.0	2	0.9	1	0.5
Transposition of the great vessels	5	2.5	5	2.5	7	3.4	10	4.8	5	2.2	9	4.1	9	4.2	6	2.8
Tetralogy of Fallot	6	3.0	4	2.0	10	4.9	17	8.1	16	7.2	4	1.8	8	3.7	6	2.8
Single ventricle	1	0.5	3	1.5	1	0.5	1	0.5	2	0.9	1	0.5	0	0.0	0	0.0
Ventricular septal defect	82	41.4	108	53.7	114	55.5	107	51.1	106	47.6	89	40.9	96	44.6	94	44.6
Atrial septal defect	58	29.3	51	25.4	49	23.9	67	32.0	70	31.4	95	43.6	91	42.3	78	37.0
Endocardial cushion defect	6	3.0	4	2.0	8	3.9	5	2.4	5	2.2	1	0.5	1	0.5	6	2.8
Pulmonary valve atresia/stenosis	13	6.6	21	10.4	15	7.3	30	14.3	26	11.7	16	7.4	18	8.4	20	9.5
Tricuspid valve atresia/stenosis	3	1.5	2	1.0	1	0.5	5	2.4	3	1.3	5	2.3	2	0.9	3	1.4
Ebstein's anomaly	1	0.5	0	0.0	0	0.0	0	0.0	0	0.0	1	0.5	0	0.0	0	0.0

TABLE 3. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY YEAR, HAWAII, 1986-2003

DIAGNOSIS	1994		1995		1996		1997		1998		1999		2000		2001	
	20,814		19,750		19,386		18,198		18,644		18,067		18,478		17,867	
	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
BRAIN & NERVOUS SYSTEM	**96**	**46.1**	**89**	**45.1**	**118**	**60.9**	**81**	**44.5**	**62**	**33.3**	**51**	**28.2**	**75**	**40.6**	**65**	**36.4**
Neural tube defects	27	13.0	20	10.1	37	19.1	17	9.3	12	6.4	15	8.3	15	8.1	16	9.0
Anencephaly	9	4.3	6	3.0	10	5.2	8	4.4	6	3.2	4	2.2	6	3.2	8	4.5
Spina bifida	12	5.8	11	5.6	14	7.2	9	4.9	5	2.7	9	5.0	6	3.2	5	2.8
Encephalocele	7	3.4	3	1.5	14	7.2	0	0.0	4	2.1	2	1.1	3	1.6	3	1.7
Microcephaly	18	8.6	25	12.7	21	10.8	17	9.3	11	5.9	6	3.3	19	10.3	17	9.5
Holoprosencephaly	7	3.4	4	2.0	2	1.0	0	0.0	3	1.6	3	1.7	5	2.7	2	1.1
Hydrocephaly (w/o spina bifida)	28	13.5	26	13.2	29	15.0	22	12.1	20	10.7	14	7.7	20	10.8	13	7.3
EYE, EAR, FACE, & NECK	**106**	**50.9**	**125**	**63.3**	**117**	**60.4**	**102**	**56.1**	**96**	**51.5**	**88**	**48.7**	**90**	**48.7**	**64**	**35.8**
EYE	44	21.1	49	24.8	51	26.3	38	20.9	39	20.9	26	14.4	30	16.2	19	10.6
Anophthalmia	1	0.5	0	0.0	1	0.5	2	1.1	0	0.0	2	1.1	1	0.5	1	0.6
Microphthalmia	6	2.9	12	6.1	7	3.6	6	3.3	3	1.6	3	1.7	3	1.6	3	1.7
Cataract	2	1.0	6	3.0	3	1.5	5	2.7	1	0.5	2	1.1	0	0.0	1	0.6
EAR	73	35.1	91	46.1	84	43.3	69	37.9	60	32.2	50	27.7	65	35.2	43	24.1
Absent auditory canal/ear	2	1.0	2	1.0	0	0.0	3	1.6	2	1.1	3	1.7	4	2.2	3	1.7
Microtia	3	1.4	7	3.5	7	3.6	9	4.9	7	3.8	6	3.3	7	3.8	4	2.2
CARDIAC & CIRCULATORY	**382**	**183.5**	**422**	**213.7**	**497**	**256.4**	**435**	**239.0**	**395**	**211.9**	**312**	**172.7**	**367**	**198.6**	**355**	**198.7**
Common truncus	1	0.5	1	0.5	3	1.5	3	1.6	3	1.6	0	0.0	0	0.0	0	0.0
Transposition of the great vessels	8	3.8	10	5.1	13	6.7	10	5.5	7	3.8	10	5.5	7	3.8	7	3.9
Tetralogy of Fallot	10	4.8	1	0.5	11	5.7	7	3.8	5	2.7	4	2.2	5	2.7	4	2.2
Single ventricle	1	0.5	4	2.0	3	1.5	1	0.5	1	0.5	3	1.7	2	1.1	0	0.0
Ventricular septal defect	103	49.5	89	45.1	102	52.6	95	52.2	77	41.3	83	45.9	66	35.7	77	43.1
Atrial septal defect	119	57.2	172	87.1	244	125.9	231	126.9	167	89.6	143	79.1	178	96.3	164	91.8
Endocardial cushion defect	6	2.9	3	1.5	11	5.7	3	1.6	5	2.7	1	0.6	7	3.8	1	0.6
Pulmonary valve atresia/stenosis	22	10.6	19	9.6	20	10.3	19	10.4	15	8.0	17	9.4	19	10.3	11	6.2
Tricuspid valve atresia/stenosis	3	1.4	6	3.0	1	0.5	2	1.1	4	2.1	1	0.6	6	3.2	2	1.1
Ebstein's anomaly	1	0.5	2	1.0	1	0.5	1	0.5	2	1.1	3	1.7	1	0.5	0	0.0

TABLE 3. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY YEAR, HAWAII, 1986-2003

DIAGNOSIS	2002		2003	
	18,182		19,033	
	N	Rate	N	Rate
BRAIN & NERVOUS SYSTEM	**72**	**39.6**	**53**	**27.8**
Neural tube defects	13	7.1	8	4.2
Anencephaly	4	2.2	2	1.1
Spina bifida	7	3.8	5	2.6
Encephalocele	2	1.1	1	0.5
Microcephaly	9	4.9	12	6.3
Holoprosencephaly	2	1.1	1	0.5
Hydrocephaly (w/o spina bifida)	19	10.4	13	6.8
EYE, EAR, FACE, & NECK	**78**	**42.9**	**82**	**43.1**
EYE	25	13.7	23	12.1
Anophthalmia	0	0.0	0	0.0
Microphthalmia	3	1.6	4	2.1
Cataract	2	1.1	3	1.6
EAR	51	28.0	56	29.4
Absent auditory canal/ear	0	0.0	2	1.1
Microtia	9	4.9	11	5.8
CARDIAC & CIRCULATORY	**359**	**197.4**	**378**	**198.6**
Common truncus	0	0.0	0	0.0
Transposition of the great vessels	11	6.0	8	4.2
Tetralogy of Fallot	8	4.4	5	2.6
Single ventricle	4	2.2	0	0.0
Ventricular septal defect	63	34.6	76	39.9
Atrial septal defect	193	106.1	201	105.6
Endocardial cushion defect	5	2.7	3	1.6
Pulmonary valve atresia/stenosis	18	9.9	12	6.3
Tricuspid valve atresia/stenosis	5	2.7	2	1.1
Ebstein's anomaly	3	1.6	2	1.1

TABLE 3. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY YEAR, HAWAII, 1986-2003

DIAGNOSIS	1986		1987		1988		1989		1990		1991		1992		1993	
	19,819		20,097		20,541		20,954		22,288		21,765		21,504		21,073	
	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Aortic valve atresia/stenosis	4	2.0	5	2.5	4	1.9	2	1.0	3	1.3	5	2.3	2	0.9	5	2.4
Mitral valve atresia/stenosis	8	4.0	3	1.5	3	1.5	1	0.5	1	0.4	6	2.8	1	0.5	1	0.5
Hypoplastic left heart syndrome	4	2.0	4	2.0	4	1.9	3	1.4	1	0.4	7	3.2	4	1.9	5	2.4
Dextrocardia	5	2.5	4	2.0	3	1.5	6	2.9	4	1.8	4	1.8	2	0.9	3	1.4
Patent ductus arteriosus*	106	53.5	150	74.6	121	58.9	105	50.1	102	45.8	138	63.4	89	41.4	113	53.6
Coarctation of the aorta	5	2.5	6	3.0	4	1.9	8	3.8	6	2.7	8	3.7	5	2.3	5	2.4
Aorta stenosis	0	0.0	0	0.0	1	0.5	1	0.5	0	0.0	0	0.0	0	0.0	0	0.0
Pulmonary artery atresia/stenosis	11	5.6	15	7.5	13	6.3	7	3.3	17	7.6	22	10.1	19	8.8	33	15.7
Total anomalous pulmonary venous return	2	1.0	1	0.5	0	0.0	1	0.5	2	0.9	3	1.4	2	0.9	3	1.4
Partial anomalous pulmonary venous	2	1.0	0	0.0	0	0.0	0	0.0	0	0.0	1	0.5	1	0.5	0	0.0
Persistent fetal circulation	31	15.6	38	18.9	21	10.2	24	11.5	17	7.6	18	8.3	10	4.7	8	3.8
RESPIRATORY	**62**	**31.3**	**51**	**25.4**	**60**	**29.2**	**59**	**28.2**	**58**	**26.0**	**54**	**24.8**	**55**	**25.6**	**50**	**23.7**
Choanal atresia/stenosis	4	2.0	3	1.5	3	1.5	4	1.9	7	3.1	1	0.5	3	1.4	1	0.5
Lung cysts	1	0.5	0	0.0	0	0.0	1	0.5	0	0.0	2	0.9	4	1.9	0	0.0
Hypoplastic lung	24	12.1	17	8.5	23	11.2	18	8.6	12	5.4	13	6.0	7	3.3	10	4.7
OROFACIAL & GASTROINTESTINAL	**124**	**62.6**	**127**	**63.2**	**151**	**73.5**	**140**	**66.8**	**145**	**65.1**	**149**	**68.5**	**131**	**60.9**	**110**	**52.2**
Oral clefts	35	17.7	45	22.4	47	22.9	33	15.7	40	17.9	42	19.3	49	22.8	41	19.5
Cleft palate (isolated)	11	5.6	14	7.0	21	10.2	6	2.9	20	9.0	14	6.4	19	8.8	19	9.0
Cleft lip (±cleft palate)	24	12.1	31	15.4	26	12.7	27	12.9	20	9.0	28	12.9	30	14.0	22	10.4
TE fistula/Esophageal atresia	9	4.5	3	1.5	7	3.4	8	3.8	6	2.7	2	0.9	1	0.5	2	0.9
Pyloric stenosis	13	6.6	16	8.0	20	9.7	30	14.3	19	8.5	16	7.4	15	7.0	18	8.5
Atresia/stenosis small intestine	2	1.0	4	2.0	4	1.9	7	3.3	7	3.1	5	2.3	5	2.3	3	1.4
Anal atresia/stenosis	10	5.0	8	4.0	11	5.4	11	5.2	13	5.8	13	6.0	8	3.7	9	4.3
Hirschsprung's disease	8	4.0	8	4.0	9	4.4	4	1.9	4	1.8	5	2.3	5	2.3	4	1.9
Malrotation	5	2.5	5	2.5	5	2.4	6	2.9	8	3.6	4	1.8	6	2.8	4	1.9
Biliary atresia	1	0.5	3	1.5	3	1.5	2	1.0	5	2.2	3	1.4	4	1.9	1	0.5
GENITAL & URINARY	**216**	**109.0**	**234**	**116.4**	**236**	**114.9**	**232**	**110.7**	**257**	**115.3**	**239**	**109.8**	**239**	**111.1**	**234**	**111.0**

TABLE 3. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY YEAR, HAWAII, 1986-2003

DIAGNOSIS	1994		1995		1996		1997		1998		1999		2000		2001	
	20,814		19,750		19,386		18,198		18,644		18,067		18,478		17,867	
	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Aortic valve atresia/stenosis	3	1.4	1	0.5	3	1.5	6	3.3	1	0.5	1	0.6	2	1.1	1	0.6
Mitral valve atresia/stenosis	0	0.0	2	1.0	3	1.5	3	1.6	1	0.5	3	1.7	0	0.0	0	0.0
Hypoplastic left heart syndrome	3	1.4	2	1.0	2	1.0	3	1.6	2	1.1	4	2.2	3	1.6	1	0.6
Dextrocardia	1	0.5	5	2.5	3	1.5	2	1.1	0	0.0	2	1.1	3	1.6	0	0.0
Patent ductus arteriosus*	144	69.2	164	83.0	191	98.5	189	103.9	204	109.4	135	74.7	191	103.4	179	100.2
Coarctation of the aorta	6	2.9	5	2.5	4	2.1	5	2.7	6	3.2	2	1.1	3	1.6	4	2.2
Aorta stenosis	0	0.0	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	48	23.1	57	28.9	61	31.5	66	36.3	44	23.6	33	18.3	49	26.5	45	25.2
Total anomalous pulmonary venous return	0	0.0	4	2.0	4	2.1	1	0.5	1	0.5	0	0.0	3	1.6	1	0.6
Partial anomalous pulmonary venous	1	0.5	3	1.5	2	1.0	0	0.0	1	0.5	2	1.1	2	1.1	2	1.1
Persistent fetal circulation	12	5.8	4	2.0	2	1.0	0	0.0	2	1.1	2	1.1	1	0.5	0	0.0
RESPIRATORY	**57**	**27.4**	**69**	**34.9**	**75**	**38.7**	**45**	**24.7**	**48**	**25.7**	**37**	**20.5**	**38**	**20.6**	**36**	**20.1**
Choanal atresia/stenosis	5	2.4	3	1.5	4	2.1	0	0.0	3	1.6	3	1.7	1	0.5	1	0.6
Lung cysts	1	0.5	1	0.5	3	1.5	2	1.1	0	0.0	1	0.6	4	2.2	2	1.1
Hypoplastic lung	6	2.9	13	6.6	9	4.6	10	5.5	4	2.1	8	4.4	8	4.3	12	6.7
OROFACIAL & GASTROINTESTINAL	**121**	**58.1**	**127**	**64.3**	**158**	**81.5**	**105**	**57.7**	**96**	**51.5**	**74**	**41.0**	**99**	**53.6**	**68**	**38.1**
Oral clefts	33	15.9	45	22.8	43	22.2	46	25.3	41	22.0	27	14.9	35	18.9	23	12.9
Cleft palate (isolated)	12	5.8	17	8.6	15	7.7	16	8.8	17	9.1	6	3.3	14	7.6	6	3.4
Cleft lip (±cleft palate)	22	10.6	28	14.2	28	14.4	30	16.5	24	12.9	21	11.6	21	11.4	17	9.5
TE fistula/Esophageal atresia	7	3.4	1	0.5	8	4.1	2	1.1	2	1.1	4	2.2	2	1.1	5	2.8
Pyloric stenosis	15	7.2	17	8.6	22	11.3	12	6.6	6	3.2	10	5.5	9	4.9	8	4.5
Atresia/stenosis small intestine	6	2.9	6	3.0	12	6.2	6	3.3	8	4.3	2	1.1	10	5.4	1	0.6
Anal atresia/stenosis	10	4.8	7	3.5	16	8.3	8	4.4	5	2.7	9	5.0	14	7.6	7	3.9
Hirschsprung's disease	5	2.4	5	2.5	4	2.1	3	1.6	7	3.8	3	1.7	4	2.2	2	1.1
Malrotation	7	3.4	11	5.6	11	5.7	5	2.7	5	2.7	5	2.8	3	1.6	4	2.2
Biliary atresia	1	0.5	1	0.5	4	2.1	1	0.5	0	0.0	2	1.1	2	1.1	0	0.0
GENITAL & URINARY	**210**	**100.9**	**245**	**124.1**	**279**	**143.9**	**190**	**104.4**	**185**	**99.2**	**144**	**79.7**	**146**	**79.0**	**175**	**97.9**

TABLE 3. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY YEAR, HAWAII, 1986-2003

DIAGNOSIS	2002		2003	
	18,182		19,033	
	N	Rate	N	Rate
Aortic valve atresia/stenosis	2	1.1	1	0.5
Mitral valve atresia/stenosis	1	0.5	0	0.0
Hypoplastic left heart syndrome	1	0.5	0	0.0
Dextrocardia	6	3.3	1	0.5
Patent ductus arteriosus*	180	99.0	202	106.1
Coarctation of the aorta	6	3.3	9	4.7
Aorta stenosis	0	0.0	0	0.0
Pulmonary artery atresia/stenosis	40	22.0	62	32.6
Total anomalous pulmonary venous return	3	1.6	1	0.5
Partial anomalous pulmonary venous	0	0.0	0	0.0
Persistent fetal circulation	2	1.1	9	4.7
RESPIRATORY	**28**	**15.4**	**27**	**14.2**
Choanal atresia/stenosis	3	1.6	1	0.5
Lung cysts	2	1.1	4	2.1
Hypoplastic lung	4	2.2	11	5.8
OROFACIAL & GASTROINTESTINAL	**72**	**39.6**	**85**	**44.7**
Oral clefts	27	14.8	28	14.7
Cleft palate (isolated)	12	6.6	10	5.3
Cleft lip (±cleft palate)	15	8.2	18	9.5
TE fistula/Esophageal atresia	1	0.5	1	0.5
Pyloric stenosis	14	7.7	12	6.3
Atresia/stenosis small intestine	3	1.6	5	2.6
Anal atresia/stenosis	5	2.7	7	3.7
Hirschsprung's disease	1	0.5	3	1.6
Malrotation	1	0.5	2	1.1
Biliary atresia	1	0.5	2	1.1
GENITAL & URINARY	**157**	**86.3**	**173**	**90.9**

TABLE 3. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY YEAR, HAWAII, 1986-2003

DIAGNOSIS	1986		1987		1988		1989		1990		1991		1992		1993	
	19,819		20,097		20,541		20,954		22,288		21,765		21,504		21,073	
	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Ovarian cyst	0	0.0	0	0.0	0	0.0	2	1.0	1	0.4	2	0.9	1	0.5	2	0.9
Undescended testicle*	86	43.4	87	43.3	101	49.2	77	36.7	90	40.4	77	35.4	99	46.0	93	44.1
Hypospadias/Epispadias/Chordee	43	21.7	69	34.3	53	25.8	56	26.7	62	27.8	59	27.1	58	27.0	60	28.5
Ambiguous genitalia	1	0.5	1	0.5	0	0.0	2	1.0	1	0.4	2	0.9	0	0.0	0	0.0
Absent/hypoplastic kidney	14	7.1	6	3.0	10	4.9	9	4.3	5	2.2	15	6.9	9	4.2	6	2.8
Cystic kidney	11	5.6	7	3.5	7	3.4	13	6.2	5	2.2	7	3.2	14	6.5	6	2.8
Hydronephrosis	12	6.1	15	7.5	13	6.3	27	12.9	30	13.5	31	14.2	27	12.6	26	12.3
Exstrophy bladder	1	0.5	0	0.0	0	0.0	1	0.5	0	0.0	0	0.0	2	0.9	0	0.0
Posterior urethral valves	4	2.0	0	0.0	1	0.5	2	1.0	1	0.4	2	0.9	0	0.0	1	0.5
LIMB & MUSCULOSKELETAL	**316**	**159.4**	**320**	**159.2**	**333**	**162.1**	**331**	**158.0**	**361**	**162.0**	**298**	**136.9**	**289**	**134.4**	**277**	**131.4**
Scoliosis	7	3.5	8	4.0	5	2.4	3	1.4	6	2.7	4	1.8	5	2.3	2	0.9
Hip dislocation	22	11.1	27	13.4	37	18.0	39	18.6	47	21.1	33	15.2	25	11.6	29	13.8
Clubfoot	110	55.5	88	43.8	94	45.8	63	30.1	76	34.1	60	27.6	64	29.8	61	28.9
Polydactyly	25	12.6	32	15.9	38	18.5	49	23.4	48	21.5	28	12.9	37	17.2	34	16.1
Syndactyly	22	11.1	13	6.5	11	5.4	15	7.2	25	11.2	20	9.2	25	11.6	18	8.5
Limb reduction deformity - upper	9	4.5	5	2.5	7	3.4	8	3.8	8	3.6	8	3.7	9	4.2	8	3.8
Limb reduction deformity - lower	1	0.5	1	0.5	4	1.9	2	1.0	5	2.2	3	1.4	4	1.9	4	1.9
Craniosynostosis	15	7.6	12	6.0	12	5.8	17	8.1	16	7.2	10	4.6	9	4.2	21	10.0
Crouzon's disease	0	0.0	0	0.0	0	0.0	0	0.0	1	0.4	0	0.0	1	0.5	0	0.0
Apert syndrome	1	0.5	0	0.0	0	0.0	1	0.5	1	0.4	0	0.0	0	0.0	1	0.5
Goldenhar syndrome	1	0.5	1	0.5	2	1.0	2	1.0	1	0.4	0	0.0	1	0.5	0	0.0
Spina bifida occulta	0	0.0	0	0.0	3	1.5	1	0.5	0	0.0	1	0.5	0	0.0	0	0.0
Dwarfism	9	4.5	5	2.5	5	2.4	5	2.4	7	3.1	2	0.9	8	3.7	8	3.8
Osteogenesis imperfecta	0	0.0	0	0.0	1	0.5	2	1.0	1	0.4	1	0.5	1	0.5	1	0.5
Diaphragmatic hernia	7	3.5	5	2.5	7	3.4	10	4.8	6	2.7	5	2.3	5	2.3	4	1.9
Abdominal wall defects	15	7.6	9	4.5	14	6.8	17	8.1	14	6.3	16	7.4	17	7.9	14	6.6
Omphalocele	6	3.0	2	1.0	4	1.9	8	3.8	3	1.3	6	2.8	6	2.8	3	1.4

TABLE 3. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY YEAR, HAWAII, 1986-2003

DIAGNOSIS	1994		1995		1996		1997		1998		1999		2000		2001	
	20,814		19,750		19,386		18,198		18,644		18,067		18,478		17,867	
	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Ovarian cyst	1	0.5	1	0.5	1	0.5	3	1.6	0	0.0	0	0.0	2	1.1	1	0.6
Undescended testicle*	80	38.4	70	35.4	90	46.4	53	29.1	64	34.3	16	8.9	12	6.5	14	7.8
Hypospadias/Epispadias/Chordee	58	27.9	74	37.5	71	36.6	53	29.1	58	31.1	68	37.6	69	37.3	87	48.7
Ambiguous genitalia	0	0.0	1	0.5	3	1.5	0	0.0	1	0.5	0	0.0	1	0.5	2	1.1
Absent/hypoplastic kidney	11	5.3	10	5.1	16	8.3	9	4.9	7	3.8	9	5.0	7	3.8	10	5.6
Cystic kidney	6	2.9	13	6.6	11	5.7	10	5.5	6	3.2	11	6.1	8	4.3	8	4.5
Hydronephrosis	18	8.6	27	13.7	25	12.9	23	12.6	27	14.5	19	10.5	16	8.7	34	19.0
Exstrophy bladder	0	0.0	0	0.0	1	0.5	0	0.0	2	1.1	2	1.1	0	0.0	0	0.0
Posterior urethral valves	0	0.0	5	2.5	2	1.0	0	0.0	0	0.0	1	0.6	1	0.5	0	0.0
LIMB & MUSCULOSKELETAL	**284**	**136.4**	**264**	**133.7**	**269**	**138.8**	**232**	**127.5**	**202**	**108.3**	**193**	**106.8**	**172**	**93.1**	**167**	**93.5**
Scoliosis	10	4.8	3	1.5	6	3.1	8	4.4	3	1.6	6	3.3	5	2.7	1	0.6
Hip dislocation	15	7.2	13	6.6	6	3.1	11	6.0	10	5.4	11	6.1	11	6.0	8	4.5
Clubfoot	63	30.3	45	22.8	72	37.1	56	30.8	40	21.5	57	31.5	45	24.4	37	20.7
Polydactyly	33	15.9	37	18.7	41	21.1	37	20.3	31	16.6	26	14.4	23	12.4	31	17.4
Syndactyly	26	12.5	14	7.1	12	6.2	24	13.2	12	6.4	12	6.6	13	7.0	7	3.9
Limb reduction deformity - upper	8	3.8	3	1.5	8	4.1	5	2.7	11	5.9	4	2.2	7	3.8	3	1.7
Limb reduction deformity - lower	1	0.5	3	1.5	1	0.5	2	1.1	6	3.2	2	1.1	4	2.2	3	1.7
Craniosynostosis	24	11.5	16	8.1	6	3.1	2	1.1	4	2.1	2	1.1	2	1.1	2	1.1
Crouzon's disease	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Apert syndrome	2	1.0	1	0.5	0	0.0	1	0.5	0	0.0	1	0.6	1	0.5	1	0.6
Goldenhar syndrome	1	0.5	2	1.0	3	1.5	1	0.5	0	0.0	0	0.0	2	1.1	0	0.0
Spina bifida occulta	1	0.5	1	0.5	0	0.0	2	1.1	1	0.5	1	0.6	0	0.0	1	0.6
Dwarfism	7	3.4	8	4.1	8	4.1	4	2.2	6	3.2	3	1.7	1	0.5	5	2.8
Osteogenesis imperfecta	2	1.0	0	0.0	1	0.5	1	0.5	2	1.1	0	0.0	1	0.5	0	0.0
Diaphragmatic hernia	2	1.0	5	2.5	4	2.1	5	2.7	3	1.6	7	3.9	3	1.6	2	1.1
Abdominal wall defects	24	11.5	26	13.2	24	12.4	24	13.2	21	11.3	14	7.7	12	6.5	11	6.2
Omphalocele	10	4.8	5	2.5	8	4.1	7	3.8	8	4.3	4	2.2	6	3.2	1	0.6

TABLE 3. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY YEAR, HAWAII, 1986-2003

DIAGNOSIS	2002		2003	
	18,182		19,033	
	N	Rate	N	Rate
Ovarian cyst	0	0.0	1	0.5
Undescended testicle*	12	6.6	17	8.9
Hypospadias/Epispadias/Chordee	76	41.8	67	35.2
Ambiguous genitalia	0	0.0	0	0.0
Absent/hypoplastic kidney	2	1.1	12	6.3
Cystic kidney	4	2.2	7	3.7
Hydronephrosis	26	14.3	40	21.0
Exstrophy bladder	0	0.0	0	0.0
Posterior urethral valves	1	0.5	1	0.5
LIMB & MUSCULOSKELETAL	**160**	**88.0**	**170**	**89.3**
Scoliosis	3	1.6	5	2.6
Hip dislocation	5	2.7	11	5.8
Clubfoot	40	22.0	56	29.4
Polydactyly	25	13.7	27	14.2
Syndactyly	8	4.4	8	4.2
Limb reduction deformity - upper	5	2.7	3	1.6
Limb reduction deformity - lower	2	1.1	3	1.6
Craniosynostosis	4	2.2	1	0.5
Crouzon's disease	0	0.0	0	0.0
Apert syndrome	0	0.0	0	0.0
Goldenhar syndrome	2	1.1	0	0.0
Spina bifida occulta	0	0.0	0	0.0
Dwarfism	3	1.6	2	1.1
Osteogenesis imperfecta	1	0.5	0	0.0
Diaphragmatic hernia	1	0.5	4	2.1
Abdominal wall defects	15	8.2	14	7.4
Omphalocele	3	1.6	5	2.6

TABLE 3. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY YEAR, HAWAII, 1986-2003

DIAGNOSIS	1986		1987		1988		1989		1990		1991		1992		1993	
	19,819		20,097		20,541		20,954		22,288		21,765		21,504		21,073	
	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Gastroschisis	6	3.0	2	1.0	3	1.5	6	2.9	4	1.8	6	2.8	5	2.3	9	4.3
Prune-belly syndrome	0	0.0	0	0.0	1	0.5	0	0.0	0	0.0	0	0.0	1	0.5	1	0.5
Poland syndrome	0	0.0	0	0.0	0	0.0	1	0.5	1	0.4	0	0.0	1	0.5	0	0.0
SKIN & INTEGUMENT	**99**	**50.0**	**161**	**80.1**	**158**	**76.9**	**193**	**92.1**	**194**	**87.0**	**162**	**74.4**	**58**	**27.0**	**63**	**29.9**
CHROMOSOME & SYNDROME	**78**	**39.4**	**89**	**44.3**	**108**	**52.6**	**106**	**50.6**	**118**	**52.9**	**148**	**68.0**	**132**	**61.4**	**115**	**54.6**
Down syndrome (trisomy 21)	29	14.6	26	12.9	39	19.0	36	17.2	28	12.6	39	17.9	32	14.9	27	12.8
Patau syndrome (trisomy 13)	1	0.5	0	0.0	2	1.0	3	1.4	6	2.7	5	2.3	8	3.7	5	2.4
Edwards syndrome (trisomy 18)	3	1.5	6	3.0	3	1.5	8	3.8	14	6.3	12	5.5	8	3.7	10	4.7
Cri du Chat syndrome	0	0.0	0	0.0	1	0.5	0	0.0	0	0.0	1	0.5	0	0.0	2	0.9
Wolff-Hirschorn syndrome	0	0.0	0	0.0	0	0.0	0	0.0	1	0.4	0	0.0	1	0.5	0	0.0
Autosomal anomalies in normal infant	8	4.0	6	3.0	8	3.9	6	2.9	9	4.0	13	6.0	11	5.1	6	2.8
Triploidy	3	1.5	1	0.5	1	0.5	1	0.5	5	2.2	2	0.9	5	2.3	4	1.9
Turner syndrome (45,X)	3	1.5	10	5.0	3	1.5	9	4.3	7	3.1	13	6.0	12	5.6	4	1.9
Klinefelter syndrome (47,XXY)	1	0.5	3	1.5	2	1.0	4	1.9	3	1.3	4	1.8	2	0.9	4	1.9
Absent spleen/Asplenia	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	1	0.5
Accessory spleen	5	2.5	1	0.5	4	1.9	2	1.0	0	0.0	4	1.8	2	0.9	2	0.9
Situs inversus	2	1.0	2	1.0	1	0.5	3	1.4	0	0.0	2	0.9	4	1.9	1	0.5
Conjoined twins	0	0.0	0	0.0	0	0.0	2	1.0	0	0.0	0	0.0	0	0.0	0	0.0
Tuberous sclerosis	0	0.0	0	0.0	1	0.5	0	0.0	2	0.9	2	0.9	0	0.0	1	0.5
Amniotic band syndrome	8	4.0	4	2.0	4	1.9	10	4.8	4	1.8	6	2.8	9	4.2	5	2.4
DiGeorge syndrome	0	0.0	1	0.5	1	0.5	2	1.0	2	0.9	0	0.0	1	0.5	1	0.5
Pierre-Robin sequence	2	1.0	4	2.0	3	1.5	0	0.0	1	0.4	1	0.5	2	0.9	4	1.9
Werdnig-Hoffman syndrome	0	0.0	1	0.5	0	0.0	1	0.5	3	1.3	0	0.0	0	0.0	1	0.5
Wolff-Parkinson-White syndrome	0	0.0	1	0.5	1	0.5	1	0.5	1	0.4	1	0.5	1	0.5	2	0.9
NEOPLASMS	**20**	**10.1**	**19**	**9.5**	**25**	**12.2**	**24**	**11.5**	**20**	**9.0**	**23**	**10.6**	**32**	**14.9**	**16**	**7.6**
CONGENITAL INFECTION	**17**	**8.6**	**28**	**13.9**	**22**	**10.7**	**27**	**12.9**	**17**	**7.6**	**16**	**7.4**	**10**	**4.7**	**12**	**5.7**
MATERNAL SUBSTANCE ABUSE	**26**	**13.1**	**34**	**16.9**	**47**	**22.9**	**146**	**69.7**	**123**	**55.2**	**113**	**51.9**	**120**	**55.8**	**168**	**79.7**

TABLE 3. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY YEAR, HAWAII, 1986-2003

DIAGNOSIS	1994		1995		1996		1997		1998		1999		2000		2001	
	20,814		19,750		19,386		18,198		18,644		18,067		18,478		17,867	
	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Gastroschisis	9	4.3	10	5.1	7	3.6	7	3.8	10	5.4	6	3.3	3	1.6	8	4.5
Prune-belly syndrome	0	0.0	0	0.0	2	1.0	1	0.5	0	0.0	0	0.0	1	0.5	2	1.1
Poland syndrome	0	0.0	1	0.5	0	0.0	0	0.0	1	0.5	0	0.0	0	0.0	0	0.0
SKIN & INTEGUMENT	**73**	**35.1**	**103**	**52.2**	**181**	**93.4**	**175**	**96.2**	**93**	**49.9**	**79**	**43.7**	**105**	**56.8**	**89**	**49.8**
CHROMOSOME & SYNDROME	**141**	**67.7**	**128**	**64.8**	**153**	**78.9**	**127**	**69.8**	**91**	**48.8**	**110**	**60.9**	**85**	**46.0**	**63**	**35.3**
Down syndrome (trisomy 21)	32	15.4	23	11.6	37	19.1	30	16.5	30	16.1	26	14.4	26	14.1	20	11.2
Patau syndrome (trisomy 13)	5	2.4	3	1.5	5	2.6	8	4.4	2	1.1	6	3.3	4	2.2	2	1.1
Edwards syndrome (trisomy 18)	13	6.2	14	7.1	18	9.3	10	5.5	5	2.7	14	7.7	8	4.3	3	1.7
Cri du Chat syndrome	1	0.5	1	0.5	0	0.0	1	0.5	0	0.0	1	0.6	0	0.0	0	0.0
Wolff-Hirschorn syndrome	2	1.0	2	1.0	0	0.0	1	0.5	0	0.0	1	0.6	0	0.0	0	0.0
Autosomal anomalies in normal infant	19	9.1	13	6.6	10	5.2	8	4.4	4	2.1	5	2.8	2	1.1	6	3.4
Triploidy	4	1.9	3	1.5	5	2.6	2	1.1	0	0.0	2	1.1	2	1.1	1	0.6
Turner syndrome (45,X)	8	3.8	11	5.6	16	8.3	6	3.3	5	2.7	13	7.2	8	4.3	4	2.2
Klinefelter syndrome (47,XXY)	2	1.0	1	0.5	6	3.1	4	2.2	3	1.6	4	2.2	3	1.6	2	1.1
Absent spleen/Asplenia	0	0.0	3	1.5	2	1.0	2	1.1	1	0.5	1	0.6	0	0.0	0	0.0
Accessory spleen	3	1.4	5	2.5	3	1.5	1	0.5	1	0.5	2	1.1	3	1.6	1	0.6
Situs inversus	2	1.0	3	1.5	4	2.1	4	2.2	2	1.1	1	0.6	2	1.1	1	0.6
Conjoined twins	0	0.0	2	1.0	2	1.0	4	2.2	0	0.0	0	0.0	0	0.0	2	1.1
Tuberous sclerosis	1	0.5	0	0.0	2	1.0	1	0.5	1	0.5	0	0.0	0	0.0	1	0.6
Amniotic band syndrome	4	1.9	2	1.0	1	0.5	4	2.2	6	3.2	2	1.1	5	2.7	1	0.6
DiGeorge syndrome	2	1.0	2	1.0	4	2.1	2	1.1	3	1.6	0	0.0	0	0.0	0	0.0
Pierre-Robin sequence	1	0.5	2	1.0	1	0.5	2	1.1	2	1.1	0	0.0	3	1.6	1	0.6
Werdnig-Hoffman syndrome	1	0.5	2	1.0	0	0.0	1	0.5	0	0.0	1	0.6	0	0.0	0	0.0
Wolff-Parkinson-White syndrome	1	0.5	1	0.5	0	0.0	1	0.5	1	0.5	2	1.1	1	0.5	2	1.1
NEOPLASMS	**29**	**13.9**	**20**	**10.1**	**32**	**16.5**	**20**	**11.0**	**10**	**5.4**	**17**	**9.4**	**11**	**6.0**	**12**	**6.7**
CONGENITAL INFECTION	**19**	**9.1**	**17**	**8.6**	**19**	**9.8**	**16**	**8.8**	**9**	**4.8**	**12**	**6.6**	**6**	**3.2**	**5**	**2.8**
MATERNAL SUBSTANCE ABUSE	**200**	**96.1**	**221**	**111.9**	**193**	**99.6**	**236**	**129.7**	**205**	**110.0**	**171**	**94.6**	**219**	**118.5**	**234**	**131.0**

TABLE 3. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY YEAR, HAWAII, 1986-2003

DIAGNOSIS	2002		2003	
	18,182		19,033	
	N	Rate	N	Rate
Gastroschisis	9	4.9	8	4.2
Prune-belly syndrome	0	0.0	0	0.0
Poland syndrome	0	0.0	0	0.0
SKIN & INTEGUMENT	**81**	**44.5**	**91**	**47.8**
CHROMOSOME & SYNDROME	**71**	**39.0**	**62**	**32.6**
Down syndrome (trisomy 21)	22	12.1	18	9.5
Patau syndrome (trisomy 13)	1	0.5	1	0.5
Edwards syndrome (trisomy 18)	7	3.8	5	2.6
Cri du Chat syndrome	0	0.0	0	0.0
Wolff-Hirschorn syndrome	0	0.0	0	0.0
Autosomal anomalies in normal infant	0	0.0	3	1.6
Triploidy	2	1.1	1	0.5
Turner syndrome (45,X)	2	1.1	6	3.2
Klinefelter syndrome (47,XXY)	0	0.0	3	1.6
Absent spleen/Asplenia	1	0.5	0	0.0
Accessory spleen	2	1.1	0	0.0
Situs inversus	2	1.1	1	0.5
Conjoined twins	0	0.0	0	0.0
Tuberous sclerosis	1	0.5	0	0.0
Amniotic band syndrome	2	1.1	0	0.0
DiGeorge syndrome	0	0.0	0	0.0
Pierre-Robin sequence	2	1.1	0	0.0
Werdnig-Hoffman syndrome	1	0.5	2	1.1
Wolff-Parkinson-White syndrome	1	0.5	1	0.5
NEOPLASMS	**14**	**7.7**	**14**	**7.4**
CONGENITAL INFECTION	**10**	**5.5**	**5**	**2.6**
MATERNAL SUBSTANCE ABUSE	**209**	**114.9**	**206**	**108.2**

TABLE 3. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY YEAR, HAWAII, 1986-2003

DIAGNOSIS	1986		1987		1988		1989		1990		1991		1992		1993	
	19,819		20,097		20,541		20,954		22,288		21,765		21,504		21,073	
	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Fetal alcohol syndrome	4	2.0	5	2.5	5	2.4	10	4.8	10	4.5	10	4.6	5	2.3	5	2.4
Illicit drugs	24	12.1	30	14.9	43	20.9	140	66.8	118	52.9	110	50.5	116	53.9	164	77.8
Marijuana	10	5.0	17	8.5	20	9.7	54	25.8	39	17.5	36	16.5	20	9.3	39	18.5
Cocaine	15	7.6	18	9.0	25	12.2	57	27.2	52	23.3	37	17.0	36	16.7	35	16.6
Methamphetamine/Amphetamine	2	1.0	1	0.5	7	3.4	63	30.1	45	20.2	58	26.6	69	32.1	113	53.6

TABLE 3. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY YEAR, HAWAII, 1986-2003

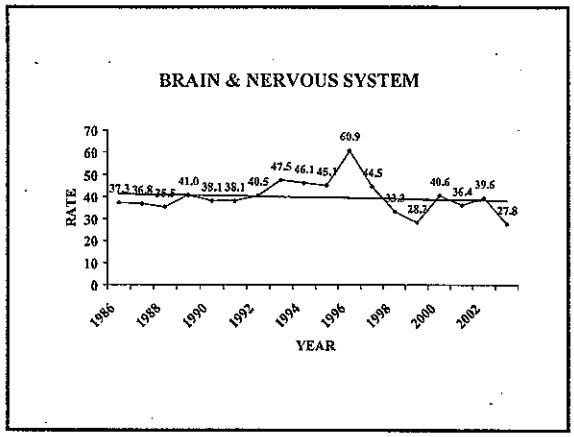
DIAGNOSIS	1994		1995		1996		1997		1998		1999		2000		2001	
	20,814		19,750		19,386		18,198		18,644		18,067		18,478		17,867	
	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Fetal alcohol syndrome	6	2.9	14	7.1	5	2.6	4	2.2	2	1.1	1	0.6	0	0.0	1	0.6
Illicit drugs	194	93.2	215	108.9	190	98.0	235	129.1	204	109.4	170	94.1	219	118.5	234	131.0
Marijuana	41	19.7	64	32.4	64	33.0	66	36.3	75	40.2	62	34.3	89	48.2	80	44.8
Cocaine	45	21.6	42	21.3	46	23.7	50	27.5	23	12.3	18	10.0	27	14.6	24	13.4
Methamphetamine/Amphetamine	126	60.5	151	76.5	113	58.3	149	81.9	140	75.1	121	67.0	157	85.0	175	97.9

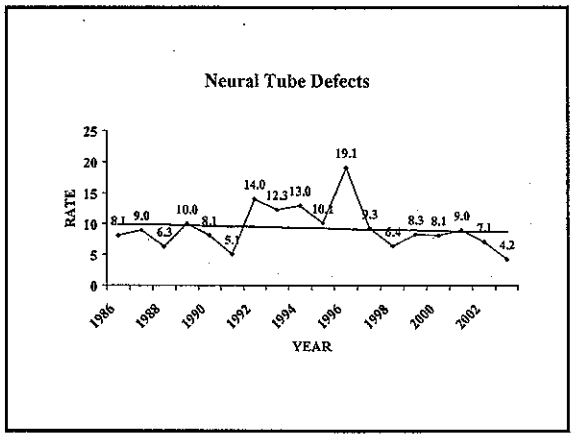
TABLE 3. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY YEAR, HAWAII, 1986-2003

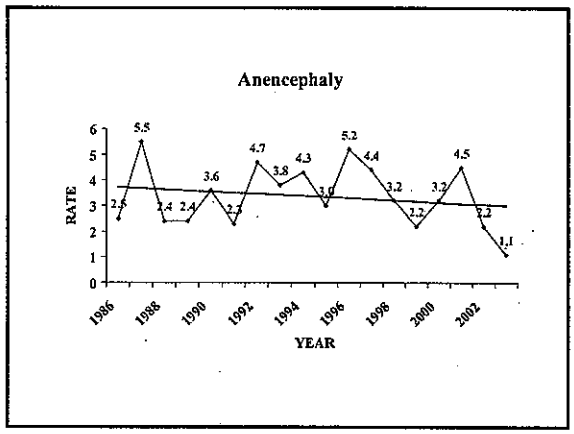
DIAGNOSIS	2002		2003	
	18,182		19,033	
	N	Rate	N	Rate
Fetal alcohol syndrome	2	1.1	0	0.0
Illicit drugs	208	114.4	206	108.2
Marijuana	68	37.4	80	42.0
Cocaine	18	9.9	8	4.2
Methamphetamine/Amphetamine	171	94.0	165	86.7

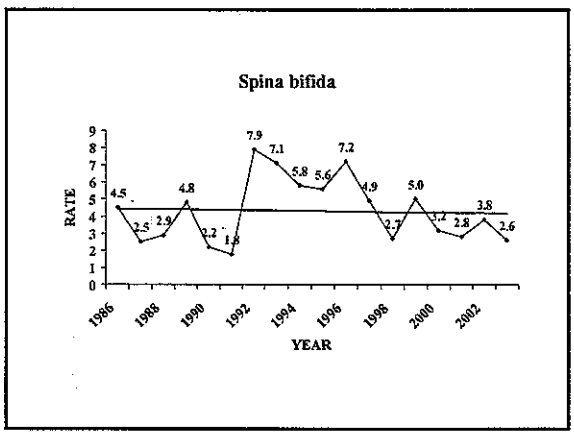
*birth weight >2499 grams.

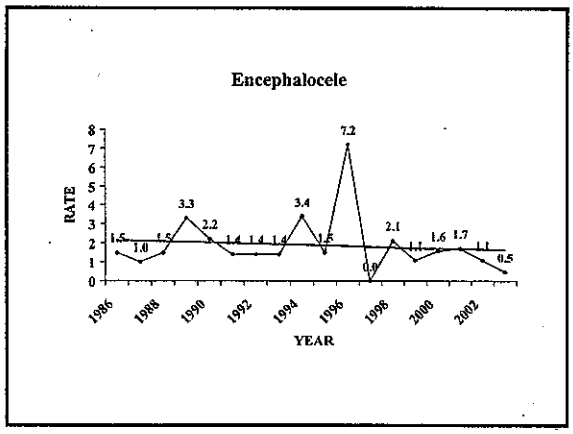
FIGURE 3. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY YEAR, HAWAII, 1986-2003











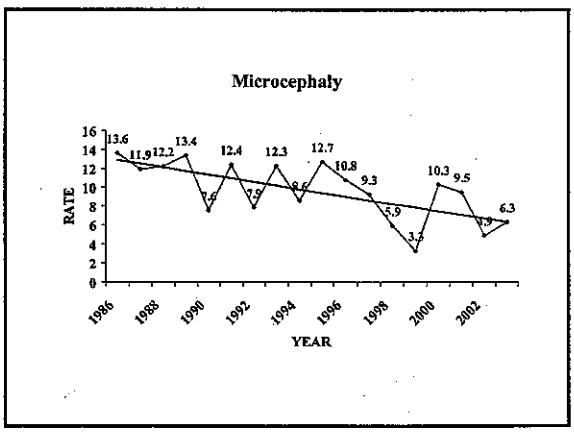
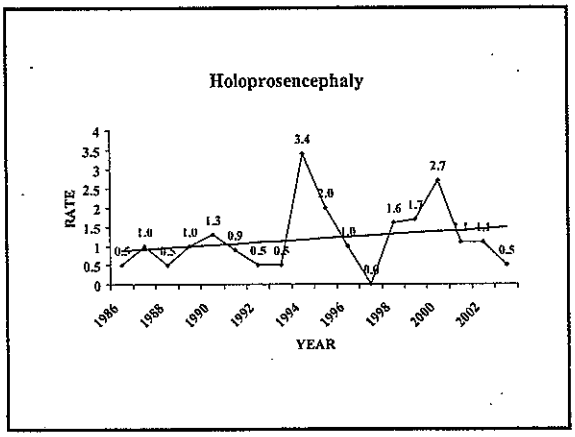
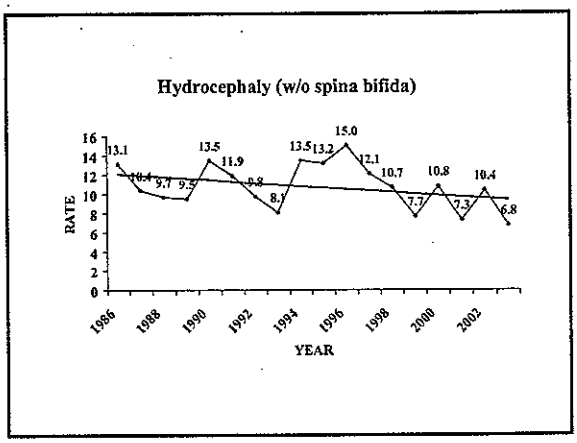
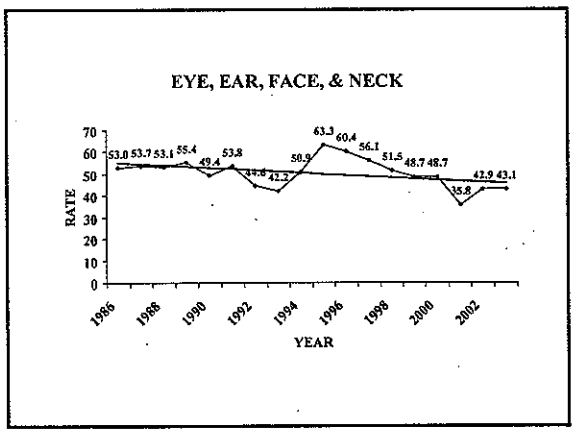
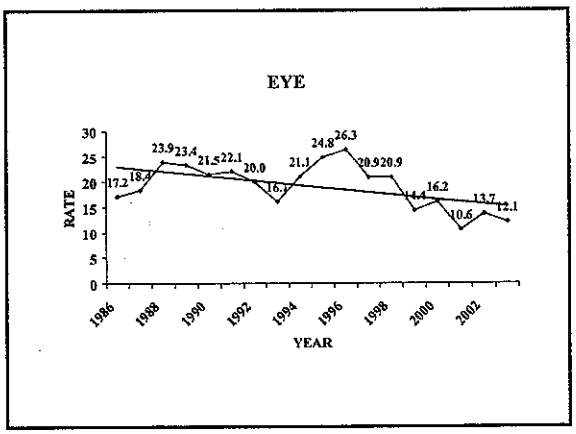


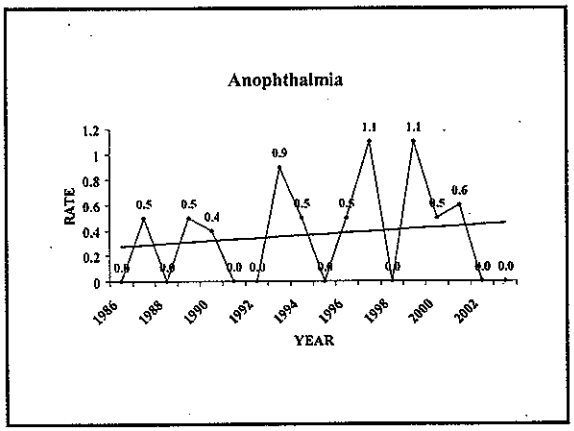
FIGURE 3. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY YEAR, HAWAII, 1986-2003

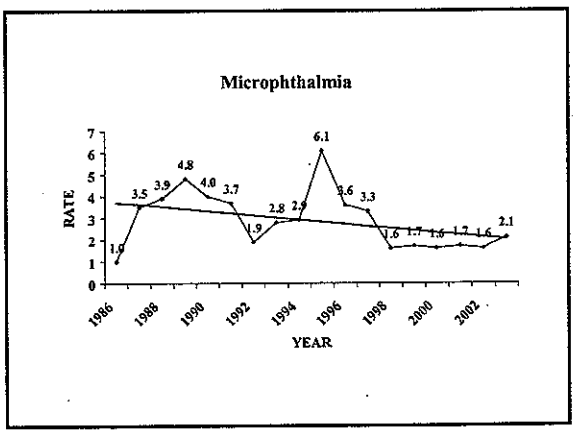












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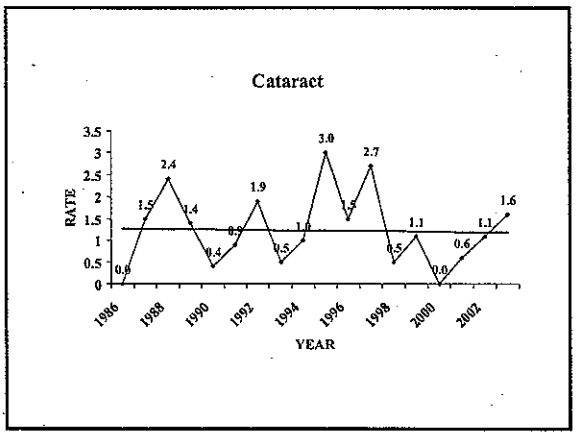

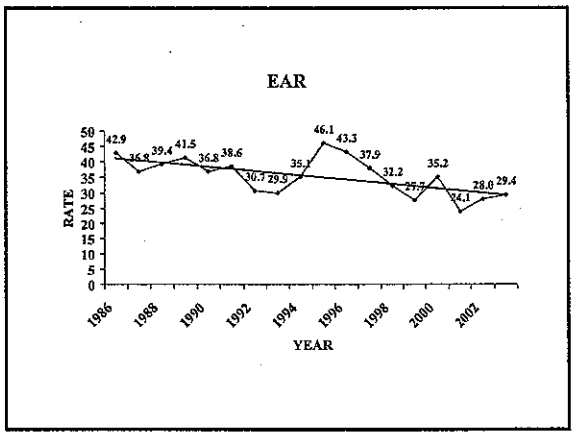

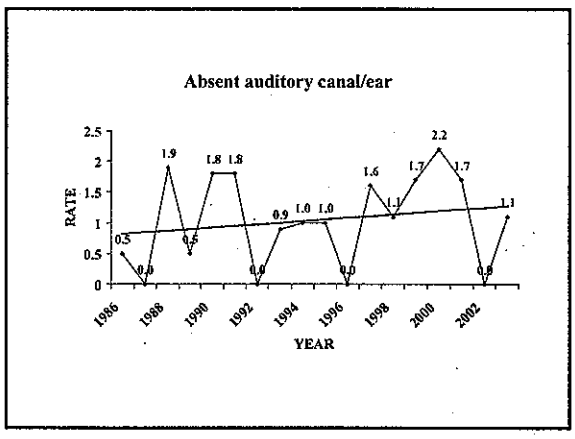

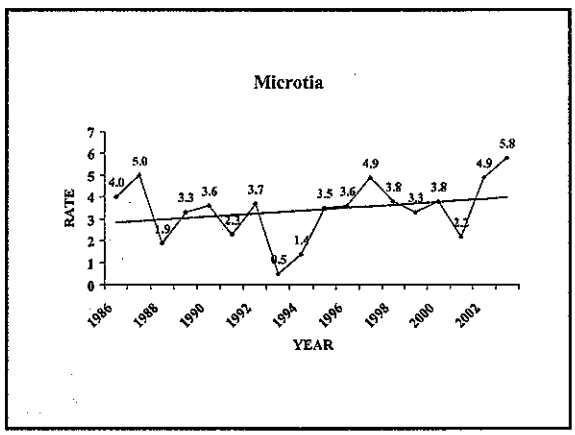

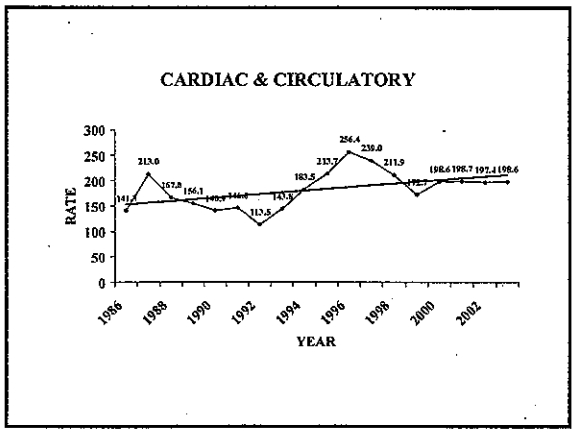

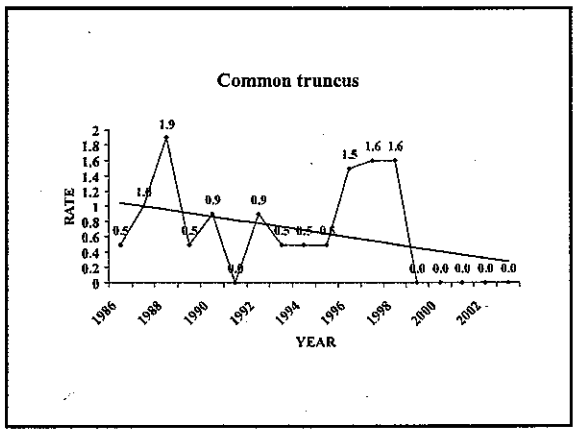

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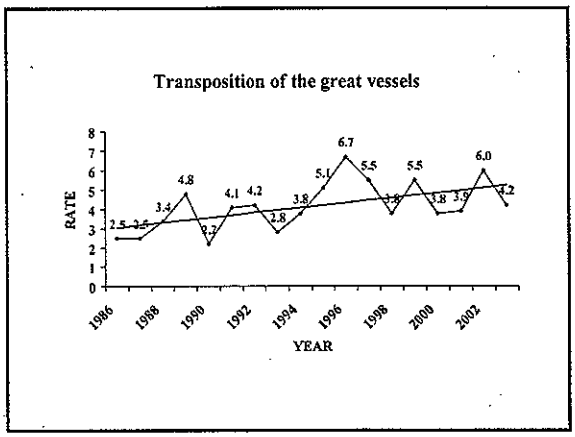

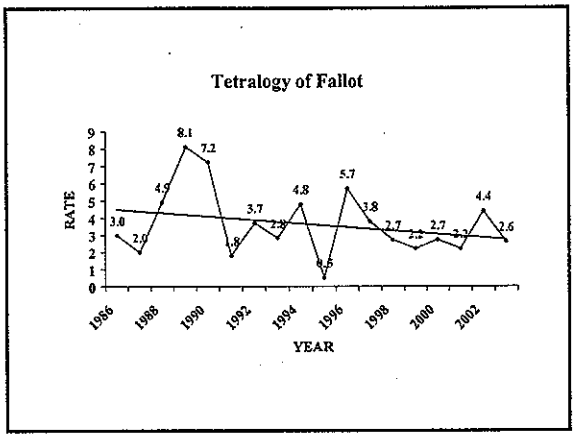

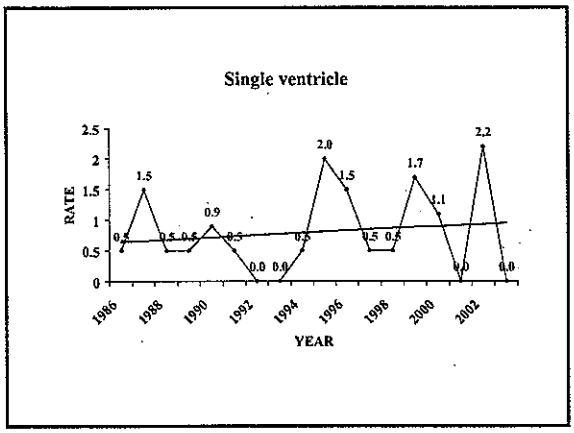

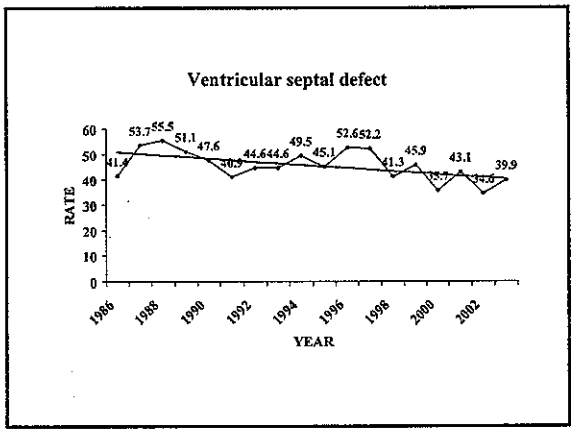

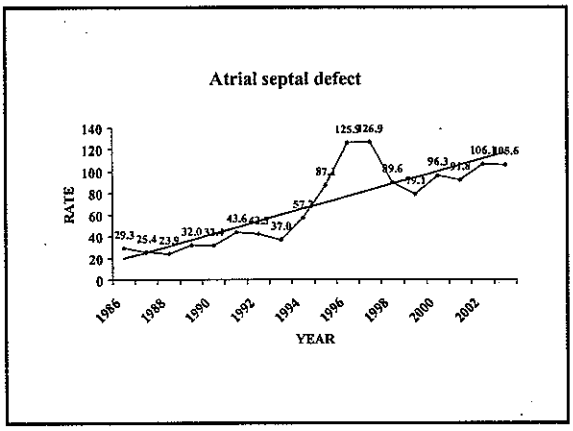

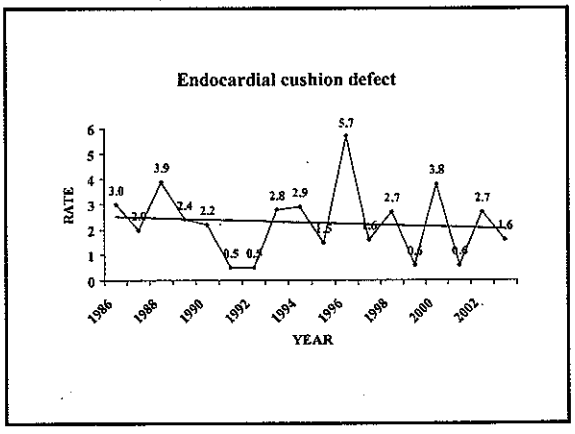

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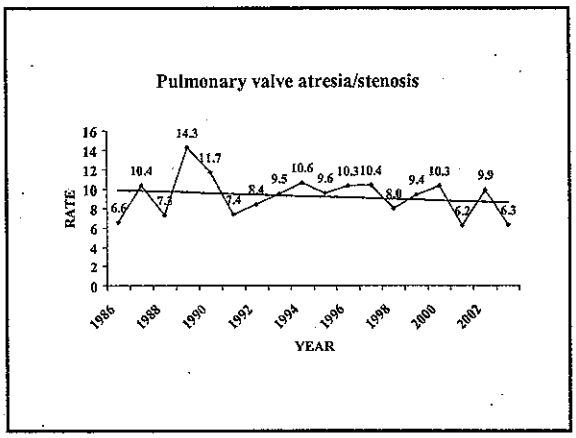

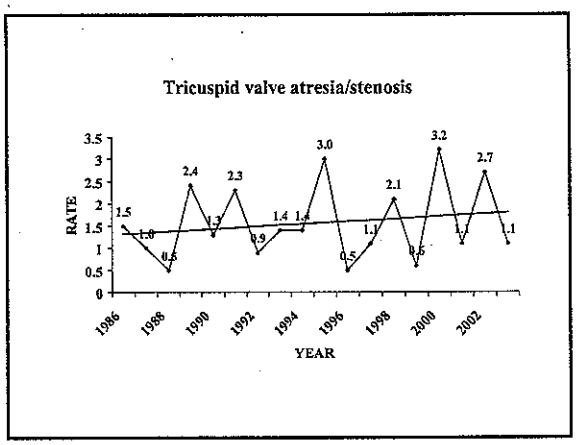

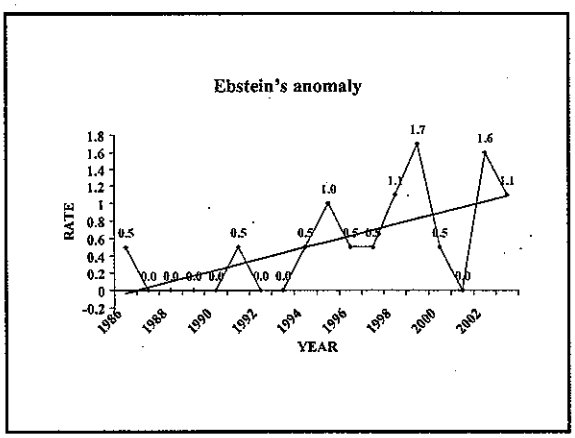

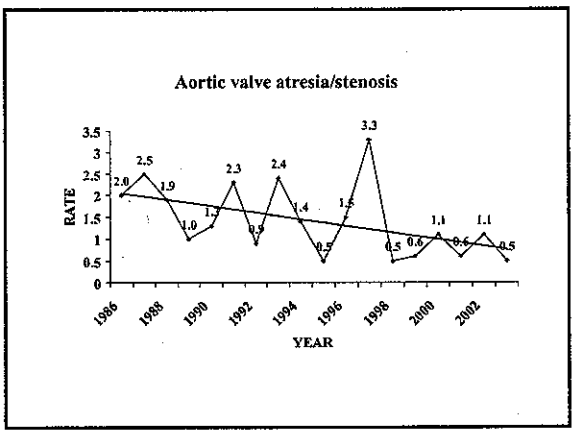

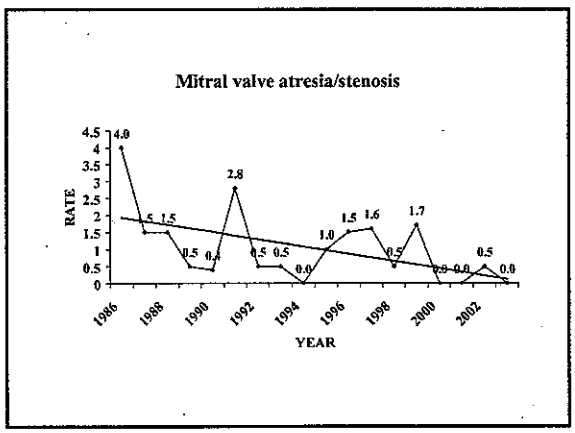

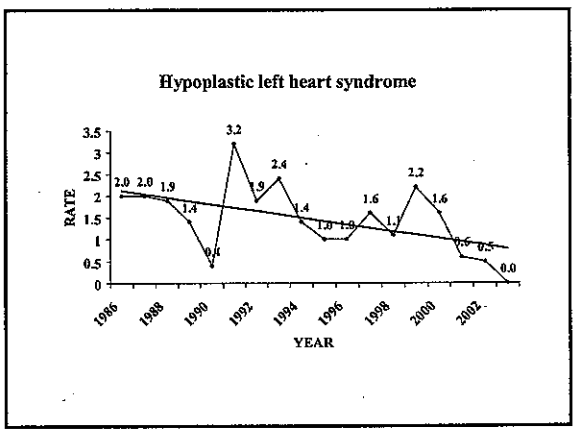

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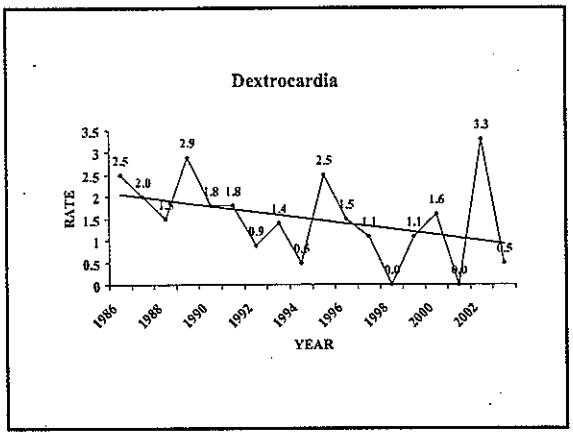
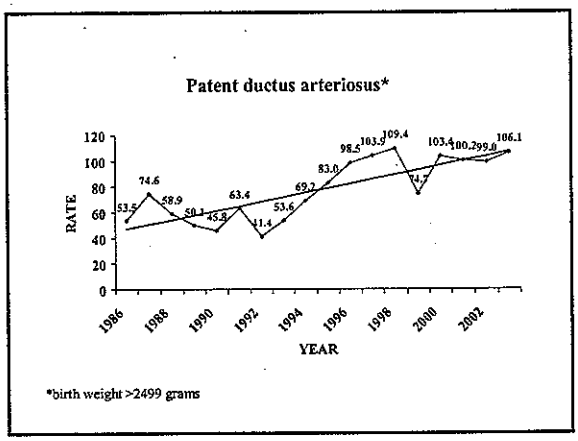
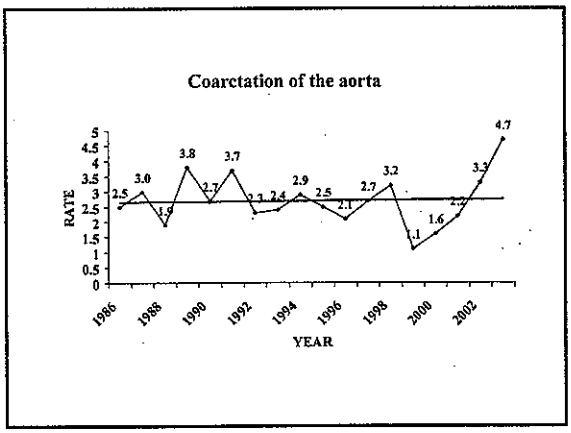
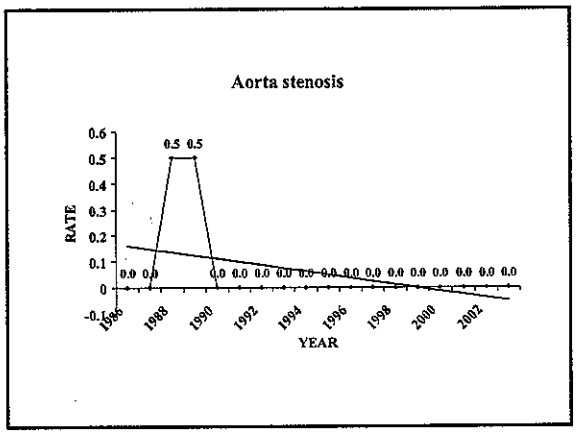
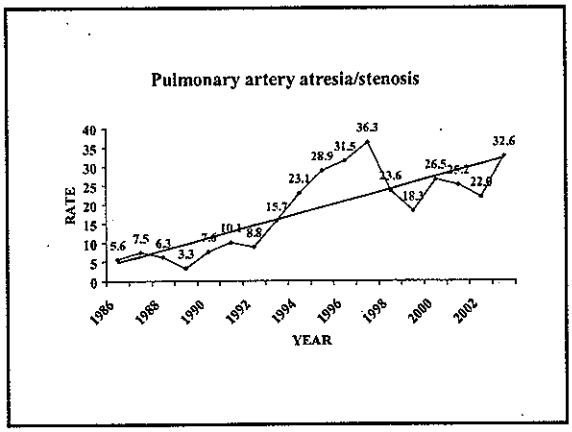










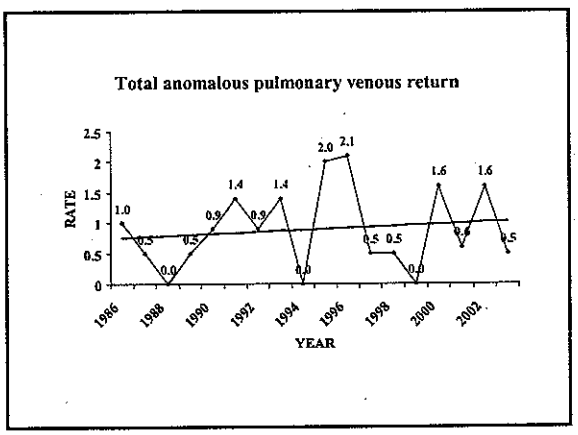
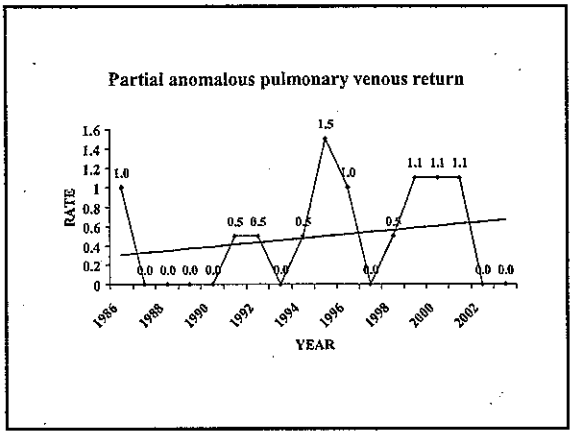
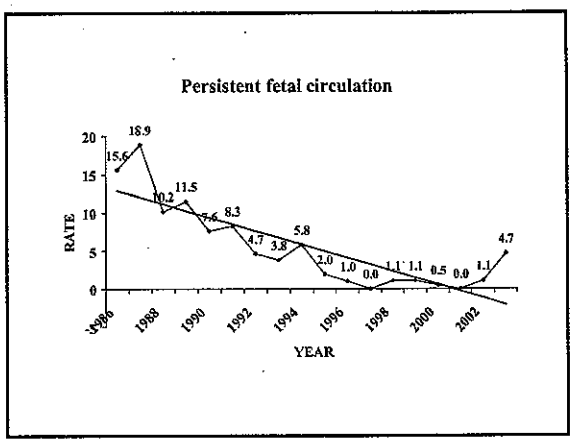
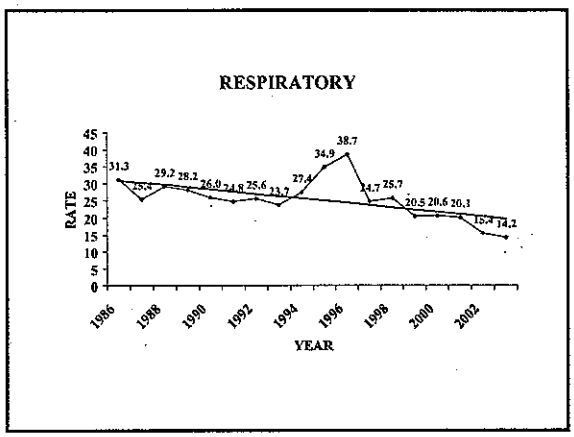
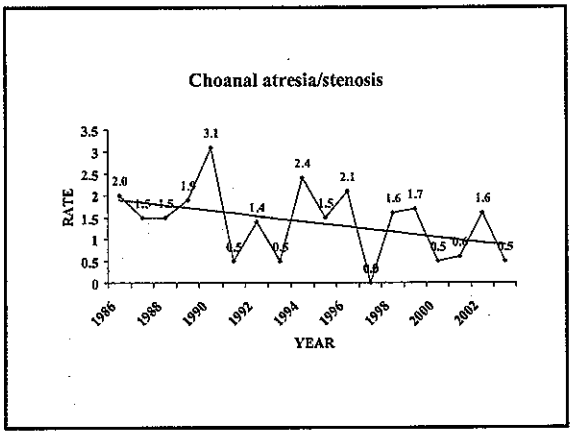


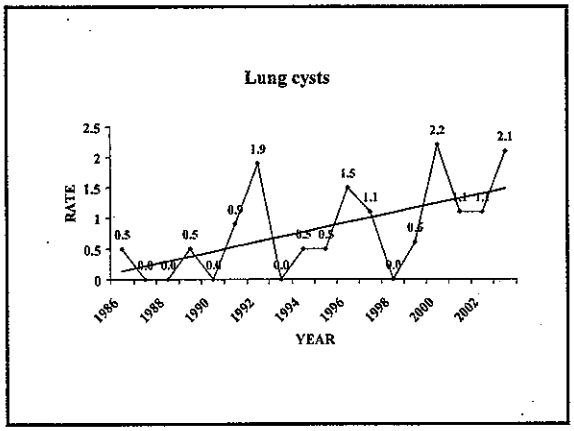
FIGURE 3. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY YEAR, HAWAII, 1986-2003

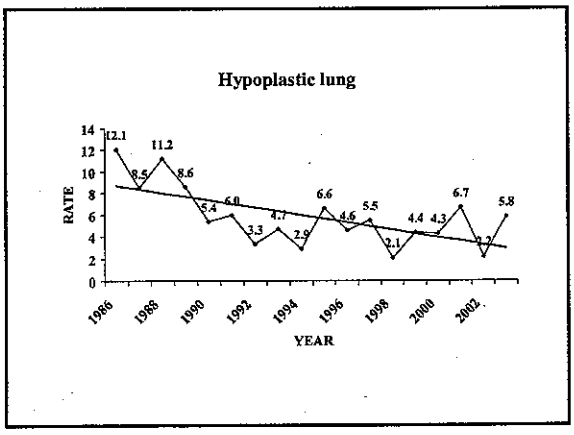












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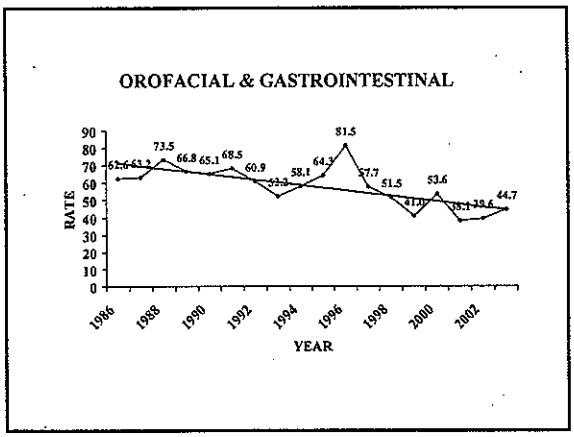

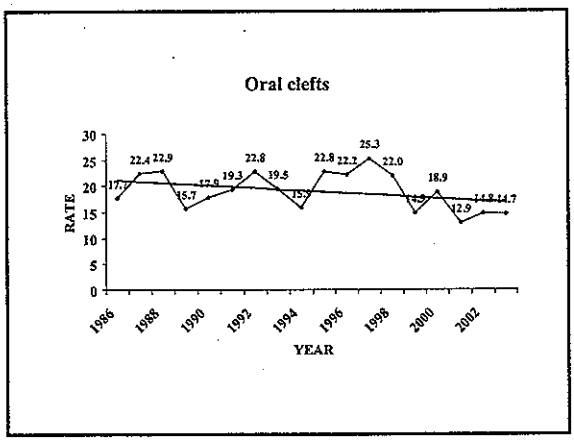

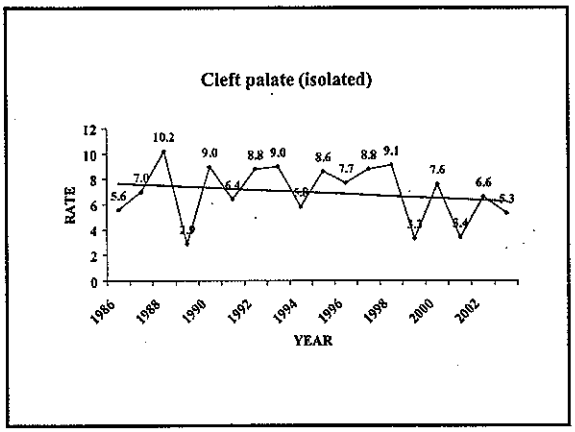

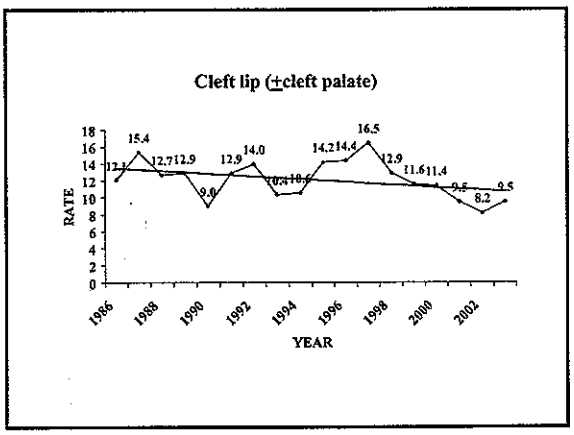

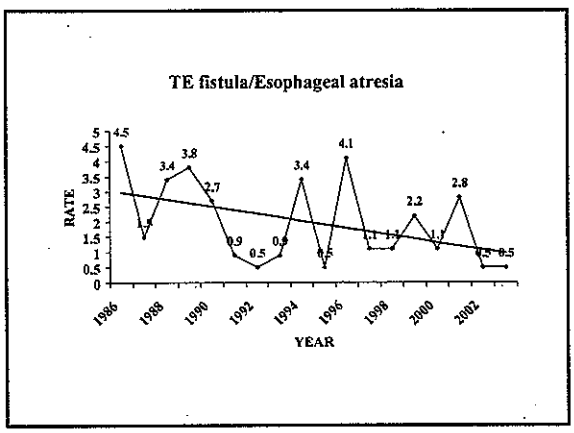

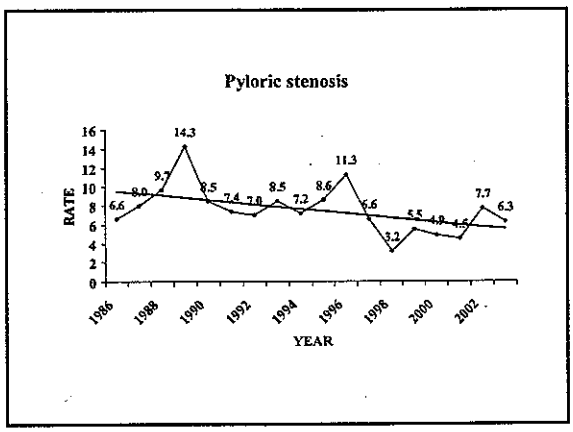

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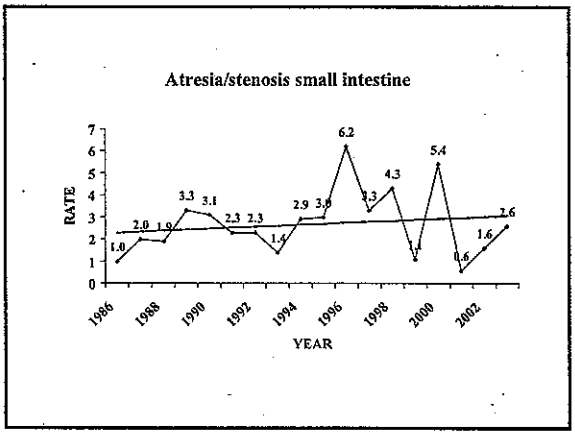

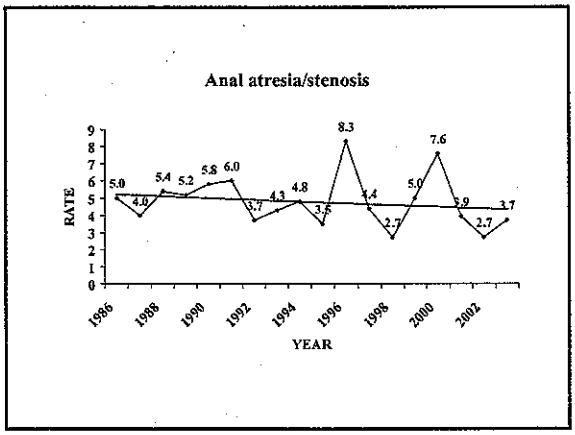

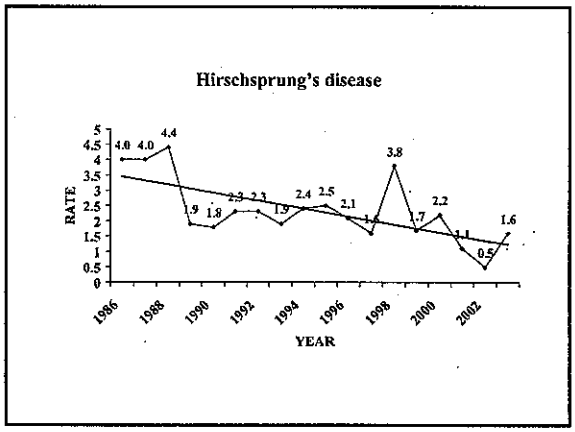

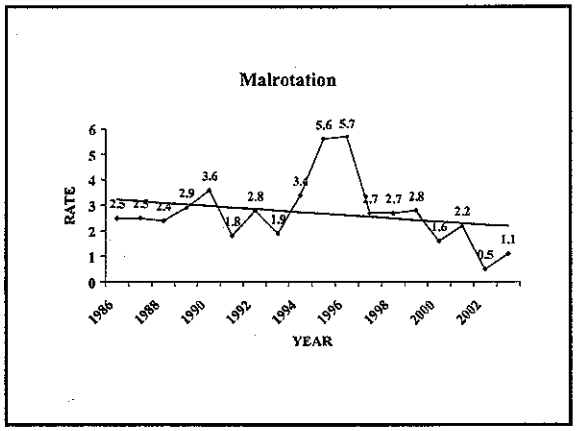

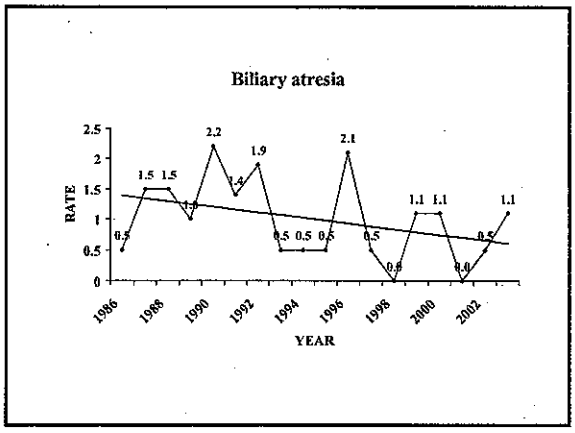

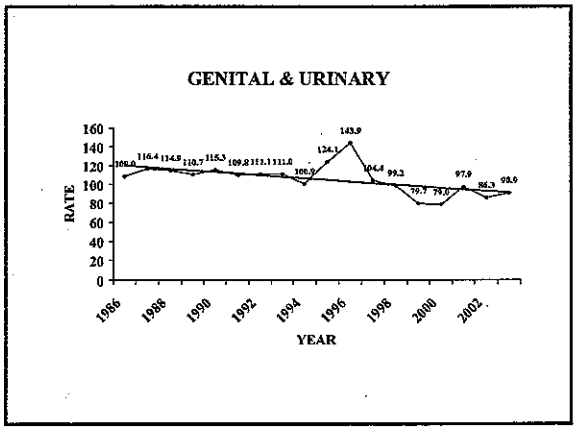

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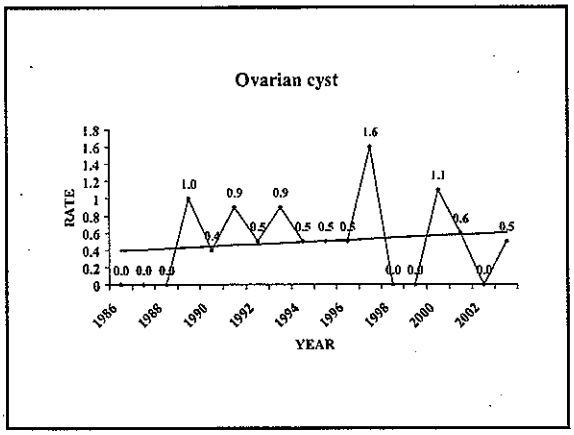
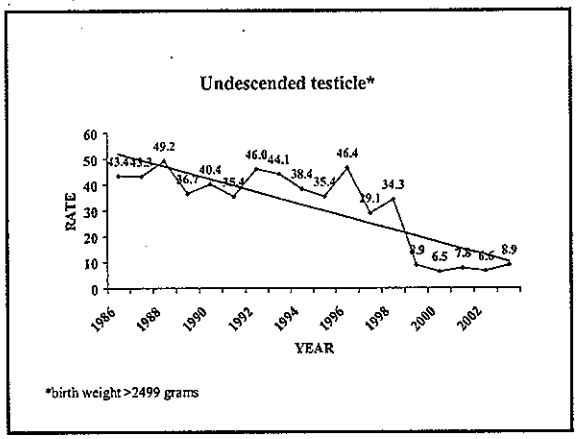
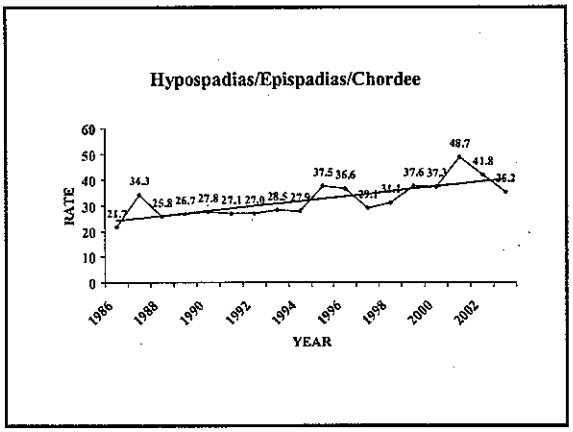
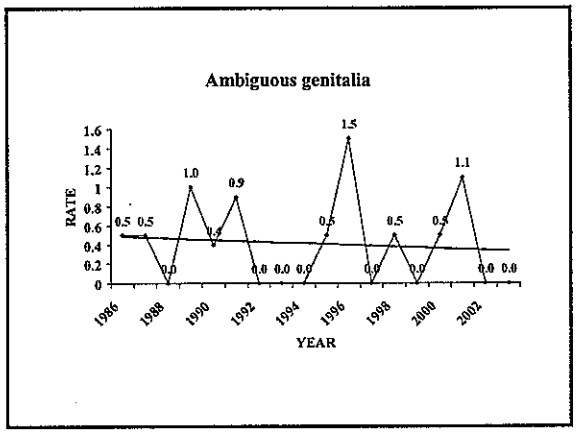
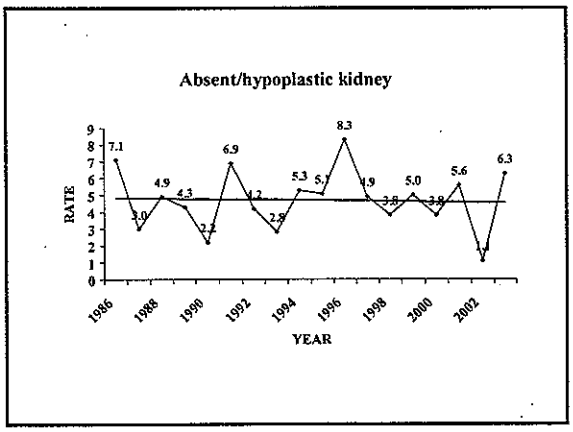










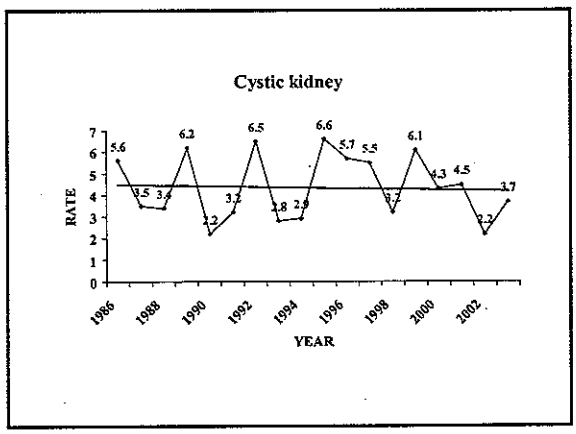
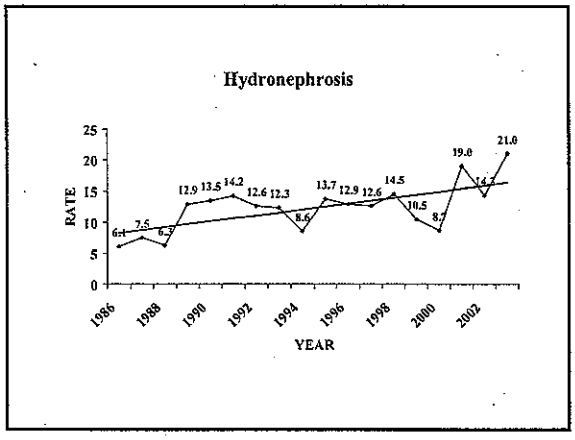
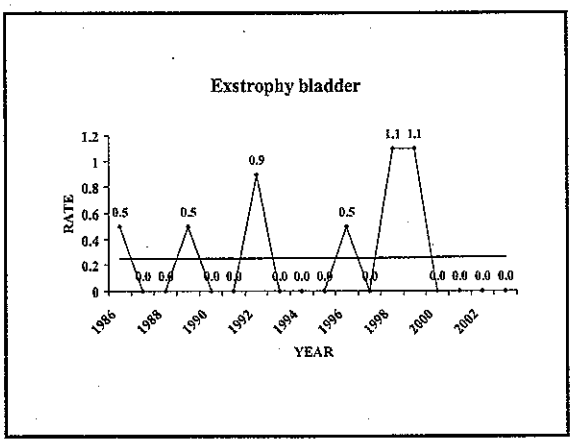
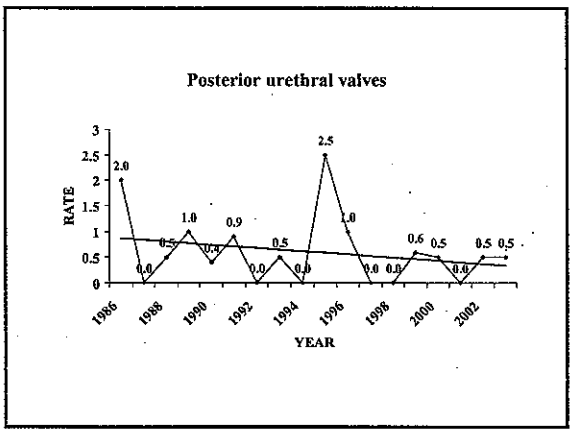
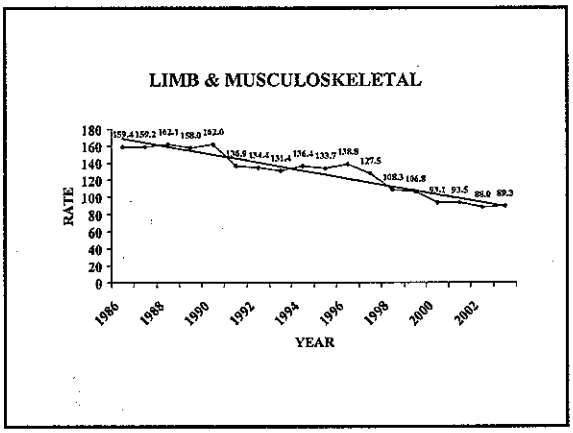


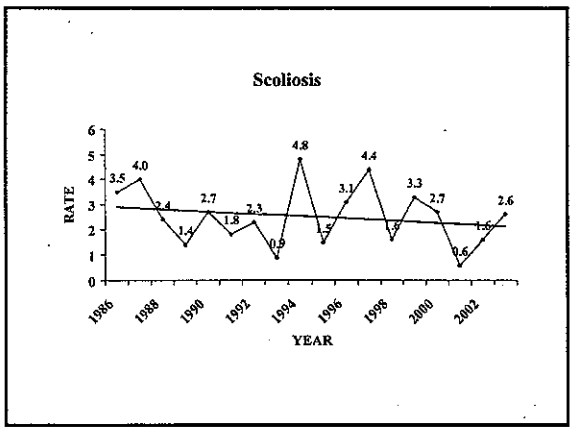
FIGURE 3. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY YEAR, HAWAII, 1986-2003

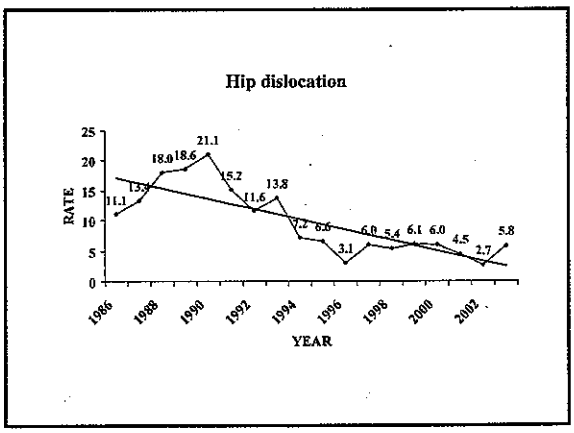












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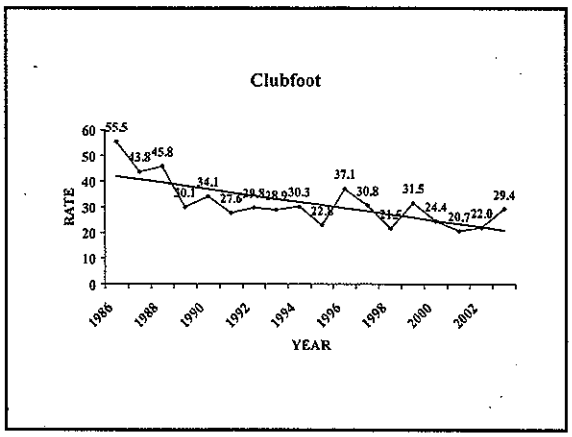

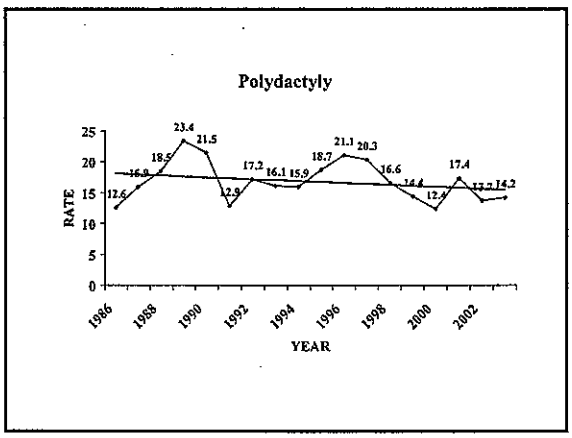

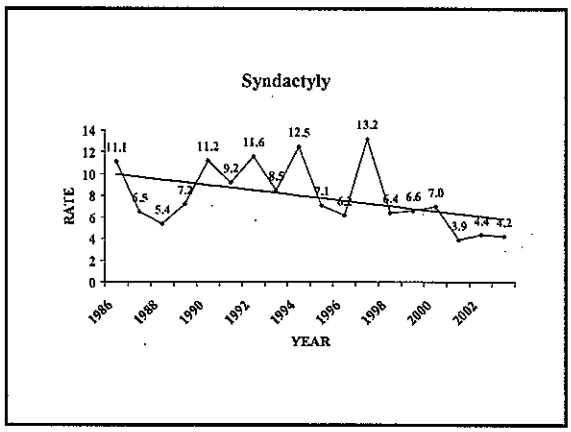

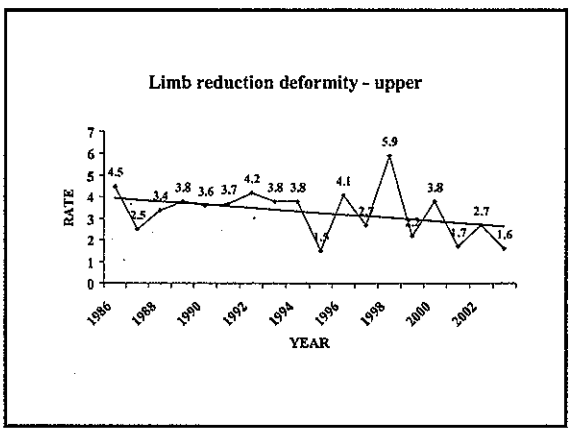

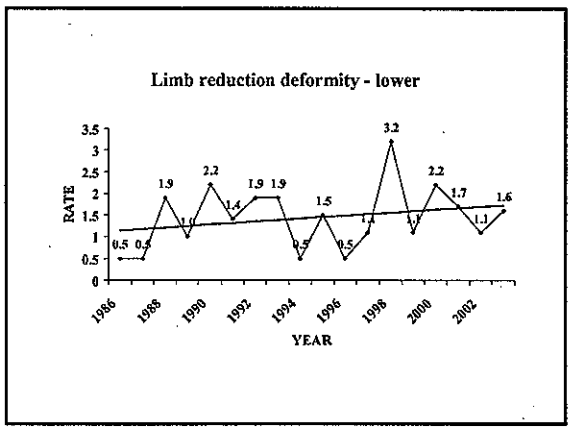

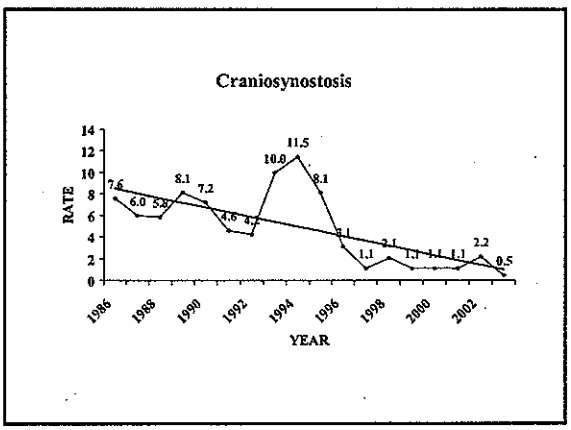

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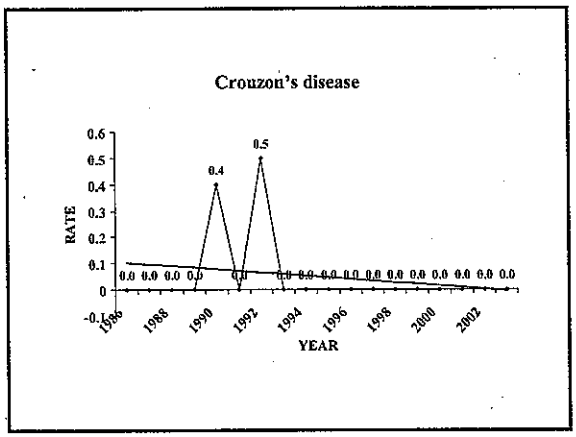

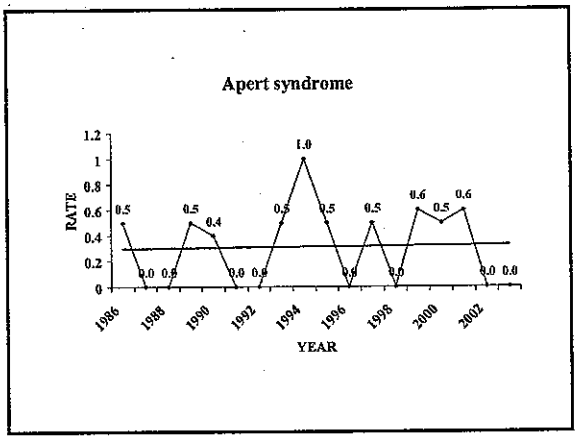

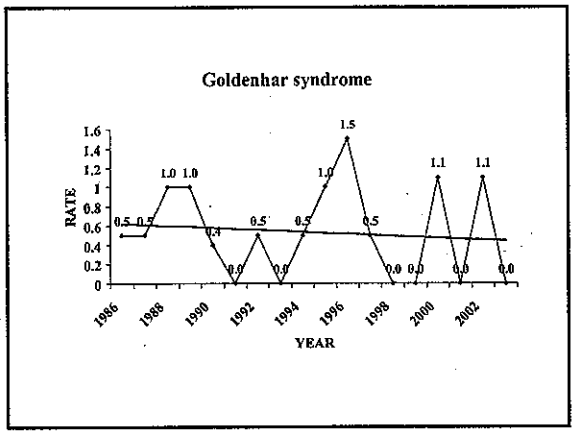

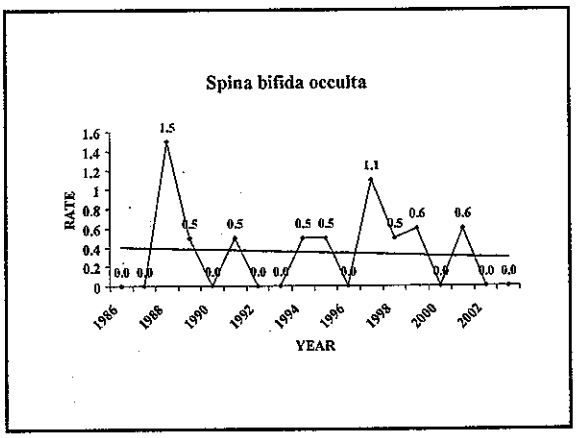

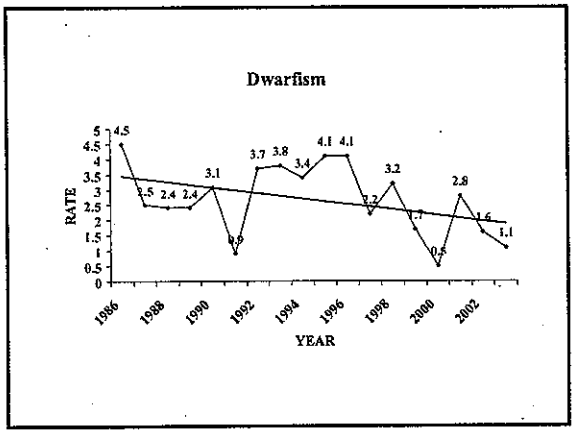

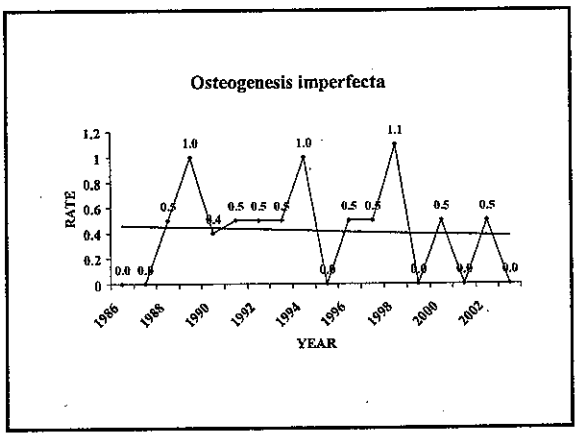

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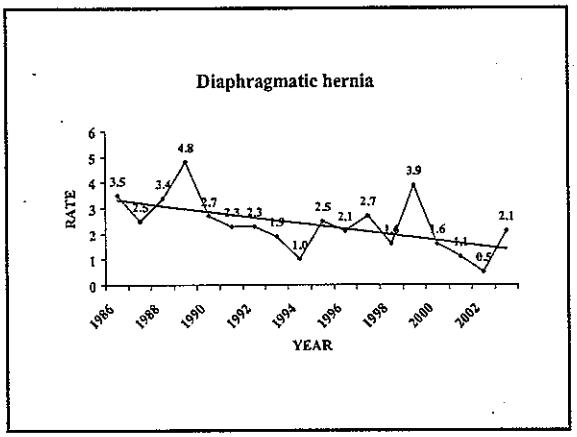

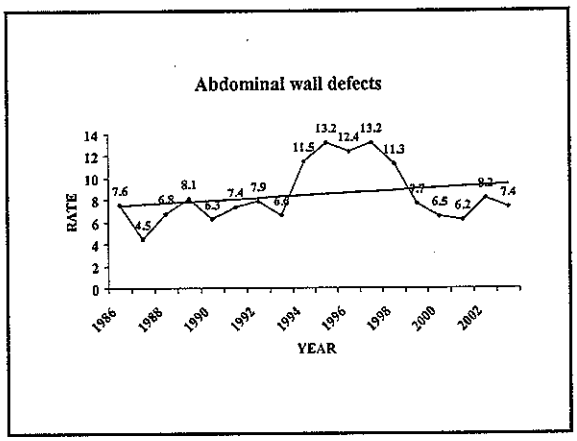

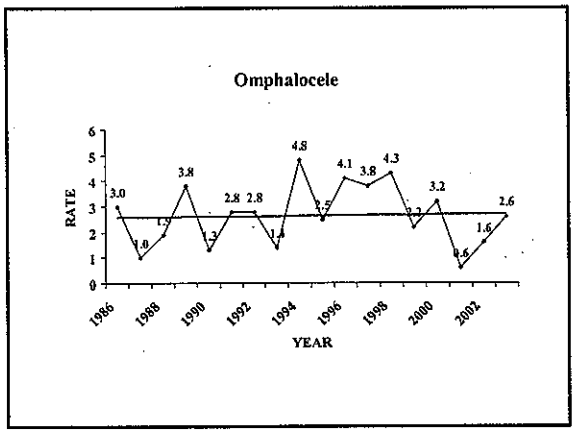

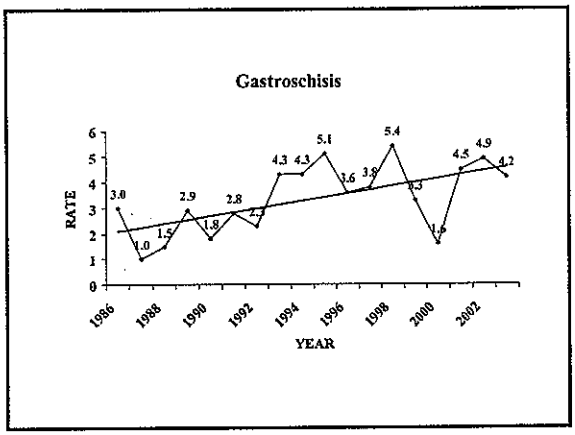

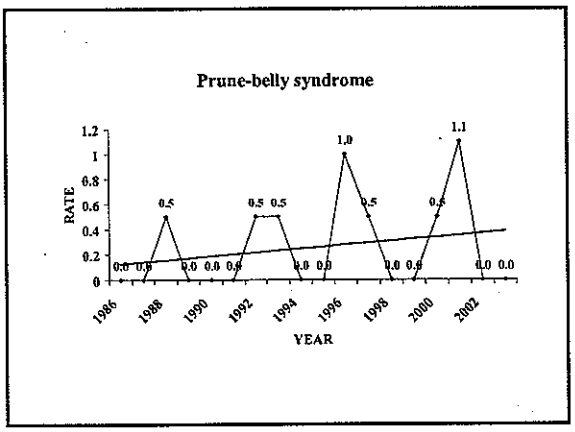

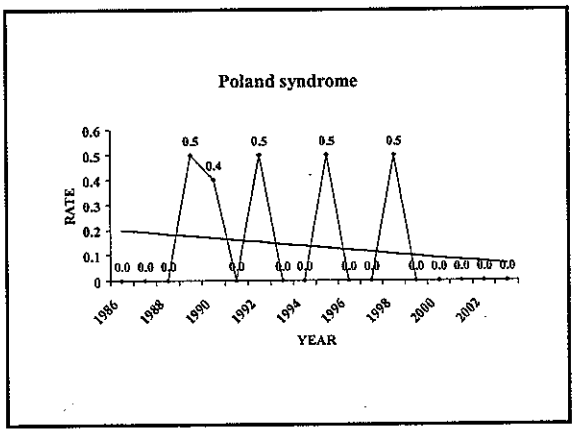

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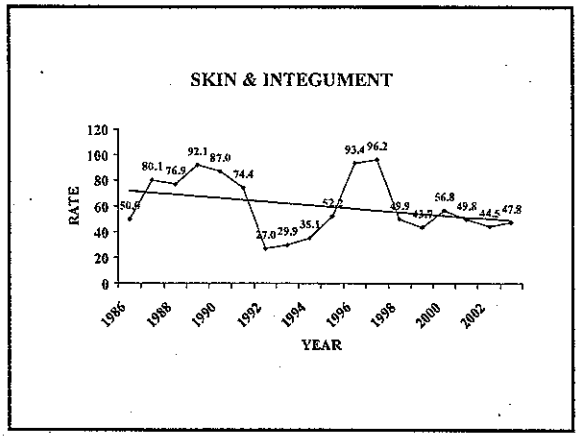
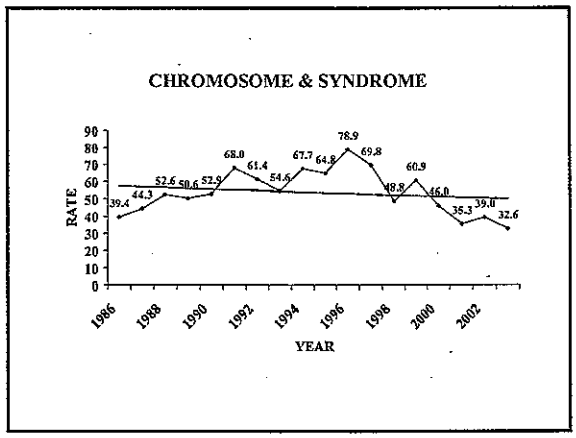
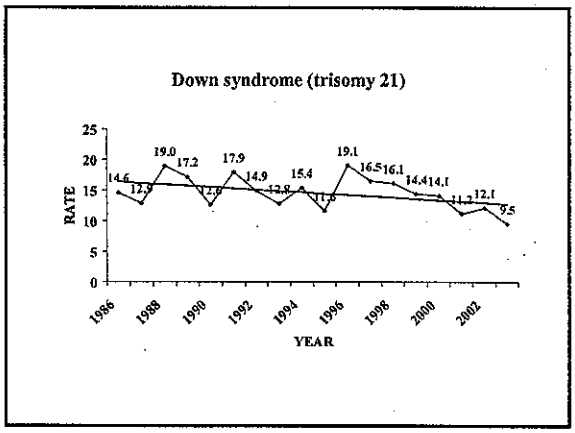
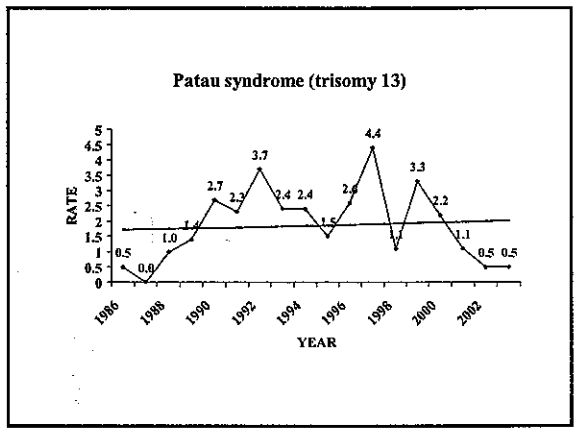
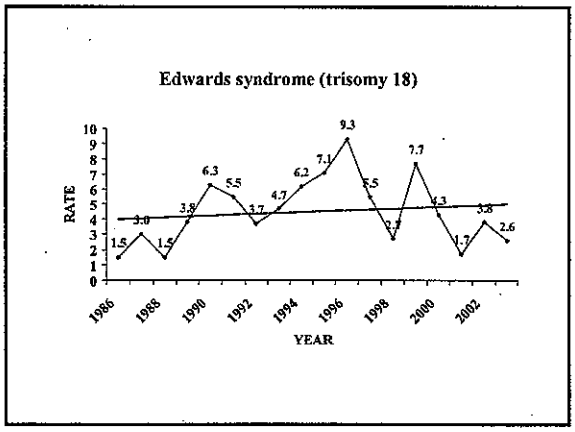










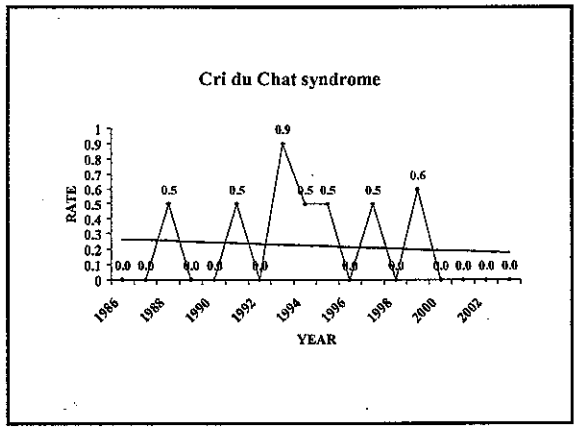
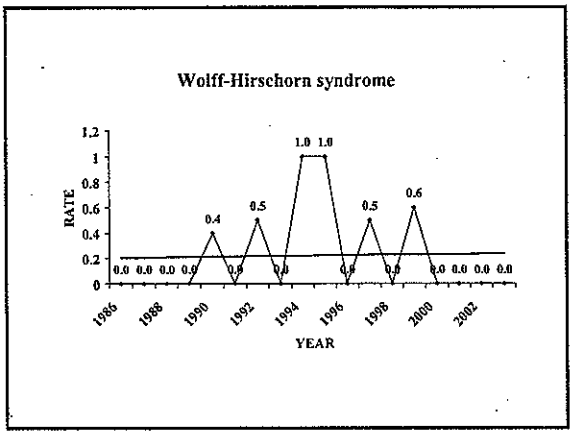
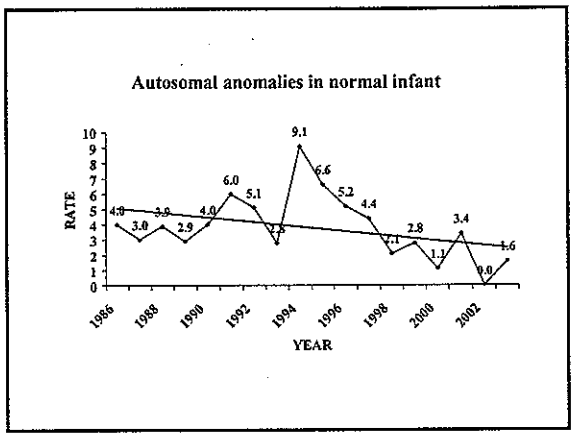
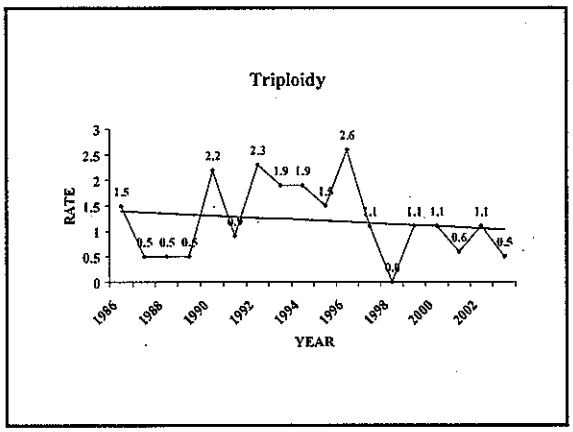
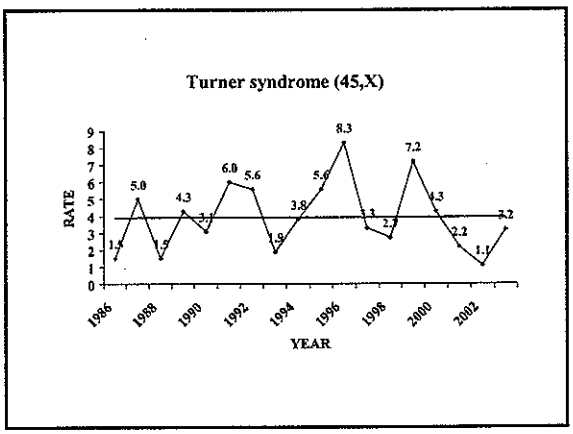


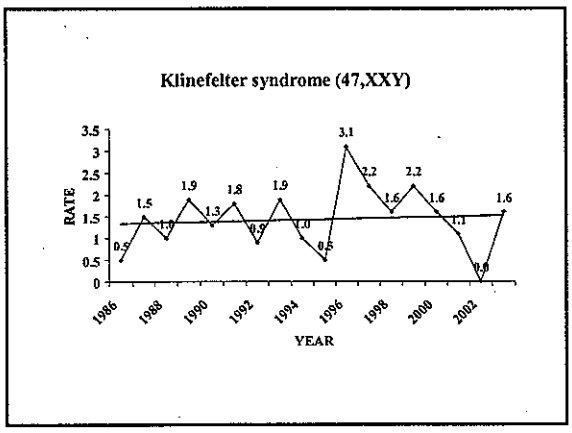
FIGURE 3. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY YEAR, HAWAII, 1986-2003

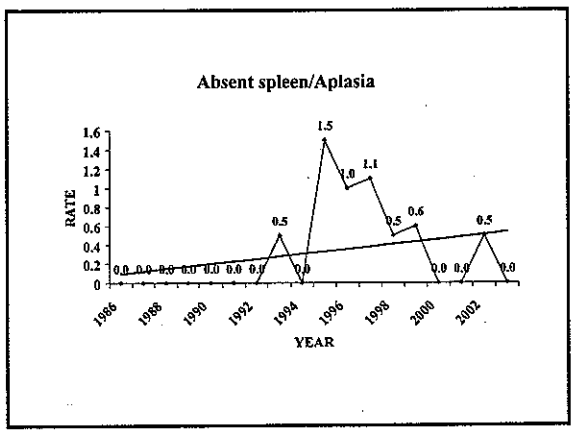












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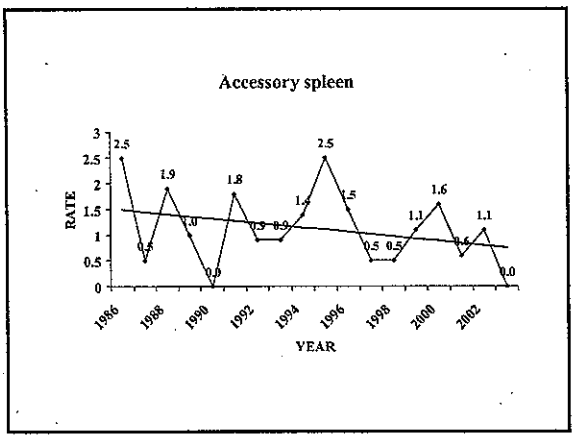

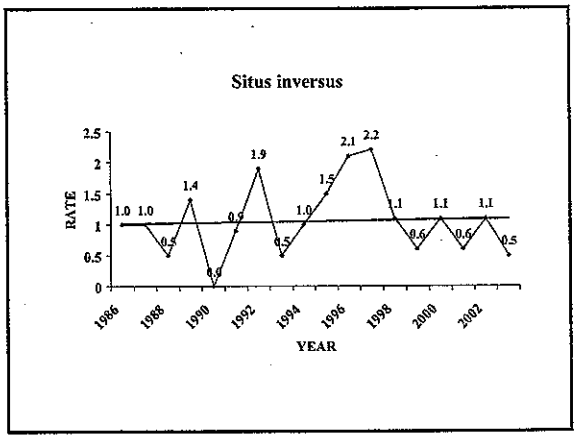

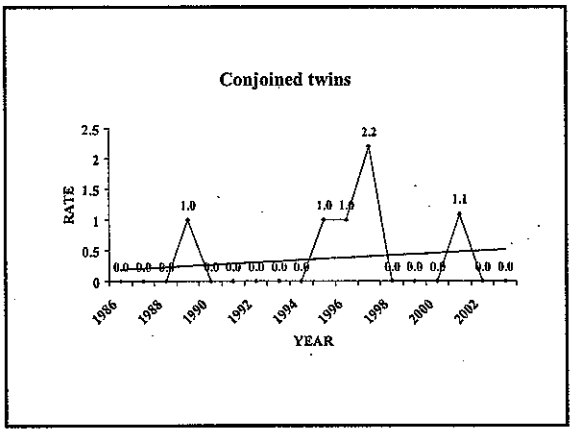

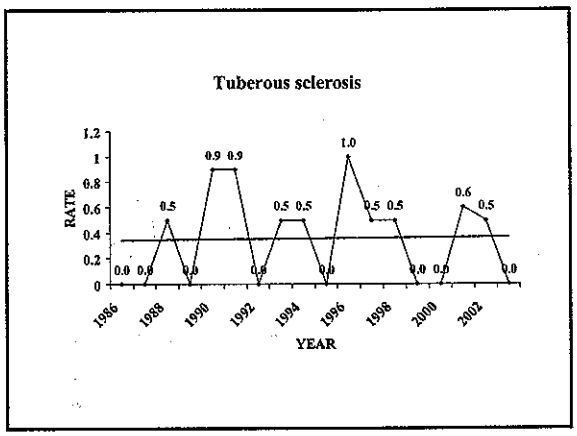

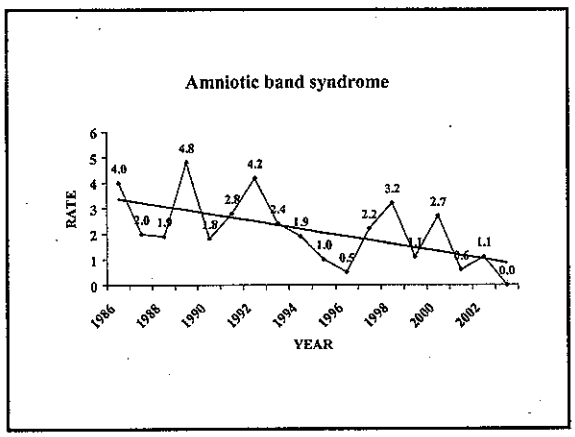

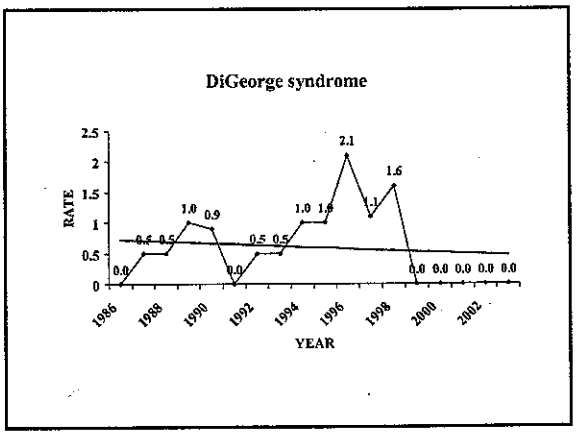

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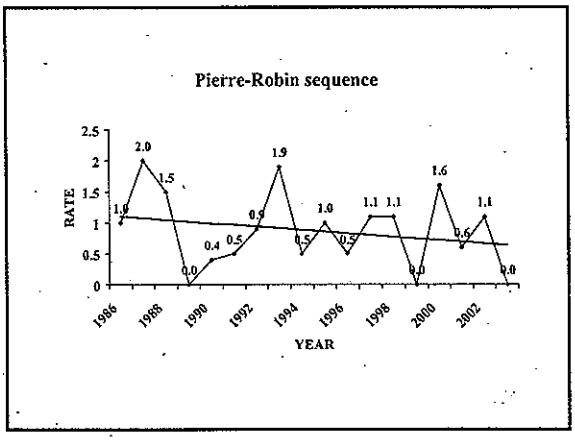

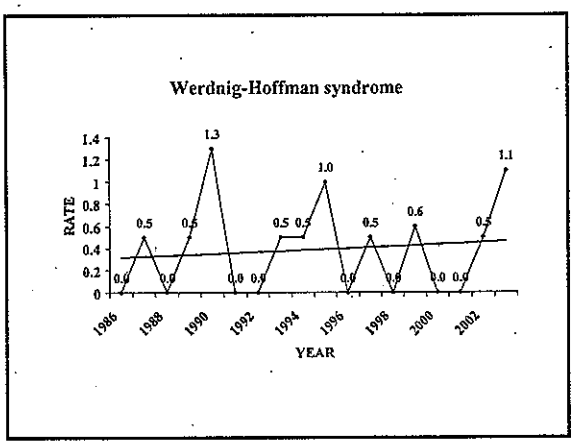

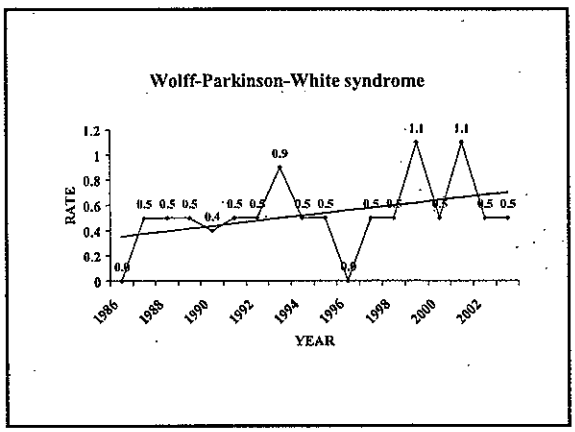

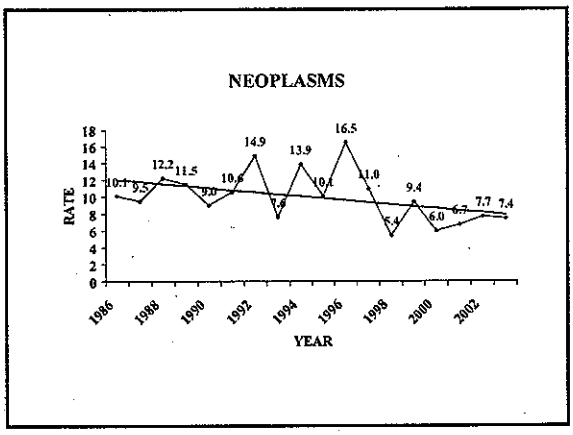

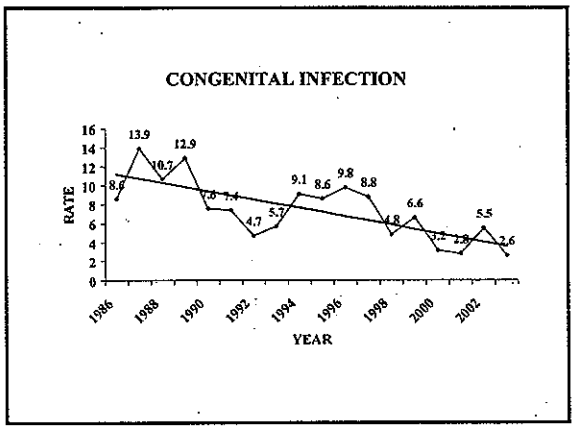

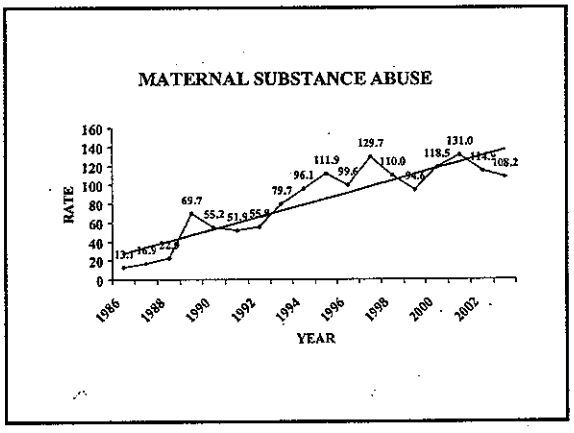

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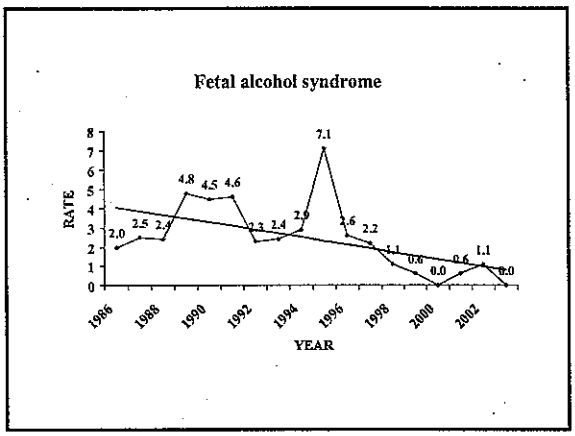
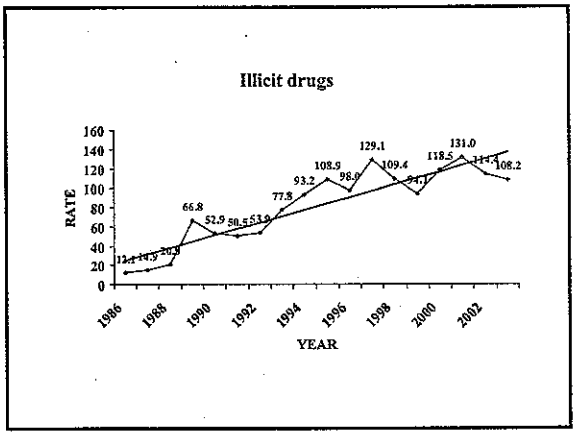
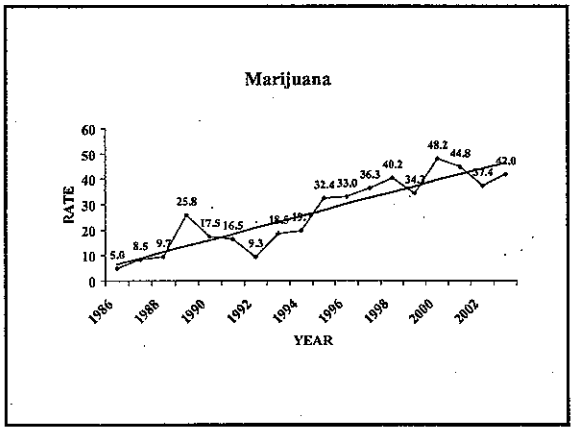
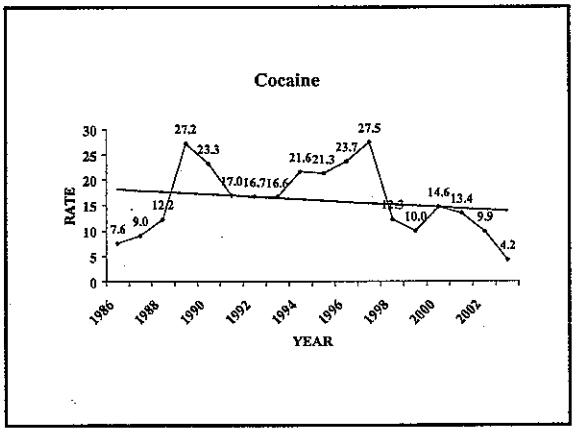








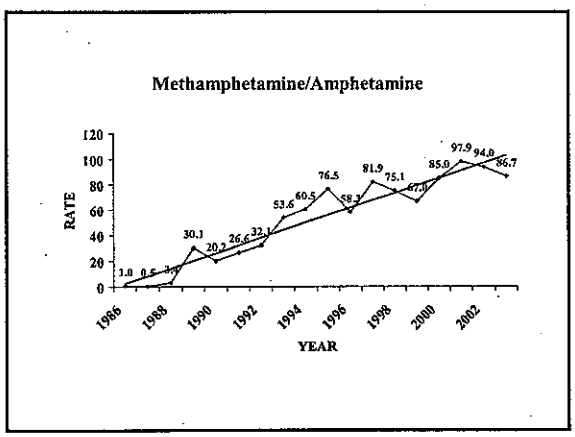


TABLE 4. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY GENDER, HAWAII, 1986-2003

DIAGNOSIS	MALE 172,222		FEMALE 161,990	
	N	Rate	N	Rate
BRAIN & NERVOUS SYSTEM	**677**	**39.3**	**679**	**41.9**
Neural tube defects	119	6.9	160	9.9
Anencephaly	40	2.3	45	2.8
Spina bifida	57	3.3	88	5.4
Encephalocele	28	1.6	30	1.9
Microcephaly	147	8.5	196	12.1
Holoprosencephaly	20	1.2	20	1.2
Hydrocephaly (w/o spina bifida)	201	11.7	173	10.7
EYE, EAR, FACE, & NECK	**992**	**57.6**	**792**	**48.9**
EYE	341	19.8	341	21.1
Anophthalmia	6	0.3	6	0.4
Microphthalmia	50	2.9	53	3.3
Cataract	20	1.2	24	1.5
EAR	727	42.2	532	32.8
Absent auditory canal/ear	20	1.2	17	1.0
Microtia	80	4.6	41	2.5
CARDIAC & CIRCULATORY	**3402**	**197.5**	**3025**	**186.7**
Common truncus	14	0.8	10	0.6
Transposition of the great vessels	87	5.1	58	3.6
Tetralogy of Fallot	62	3.6	69	4.3
Single ventricle	12	0.7	15	0.9
Ventricular septal defect	790	45.9	832	51.4
Atrial septal defect	1240	72.0	1114	68.8
Endocardial cushion defect	32	1.9	49	3.0
Pulmonary valve atresia/stenosis	172	10.0	156	9.6
Tricuspid valve atresia/stenosis	26	1.5	28	1.7
Ebstein's anomaly	7	0.4	10	0.6
Aortic valve atresia/stenosis	34	2.0	16	1.0
Mitral valve atresia/stenosis	22	1.3	15	0.9
Hypoplastic left heart syndrome	29	1.7	23	1.4
Dextrocardia	24	1.4	29	1.8
Patent ductus arteriosus*	1454	84.4	1249	77.1
Coarctation of the aorta	60	3.5	37	2.3
Aorta stenosis	1	0.1	1	0.1
Pulmonary artery atresia/stenosis	332	19.3	305	18.8
Total anomalous pulmonary venous return	6	0.3	26	1.6
Partial anomalous pulmonary venous	7	0.4	10	0.6
Persistent fetal circulation	116	6.7	85	5.2
RESPIRATORY	**512**	**29.7**	**396**	**24.4**

TABLE 4. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY GENDER, HAWAII, 1986-2003

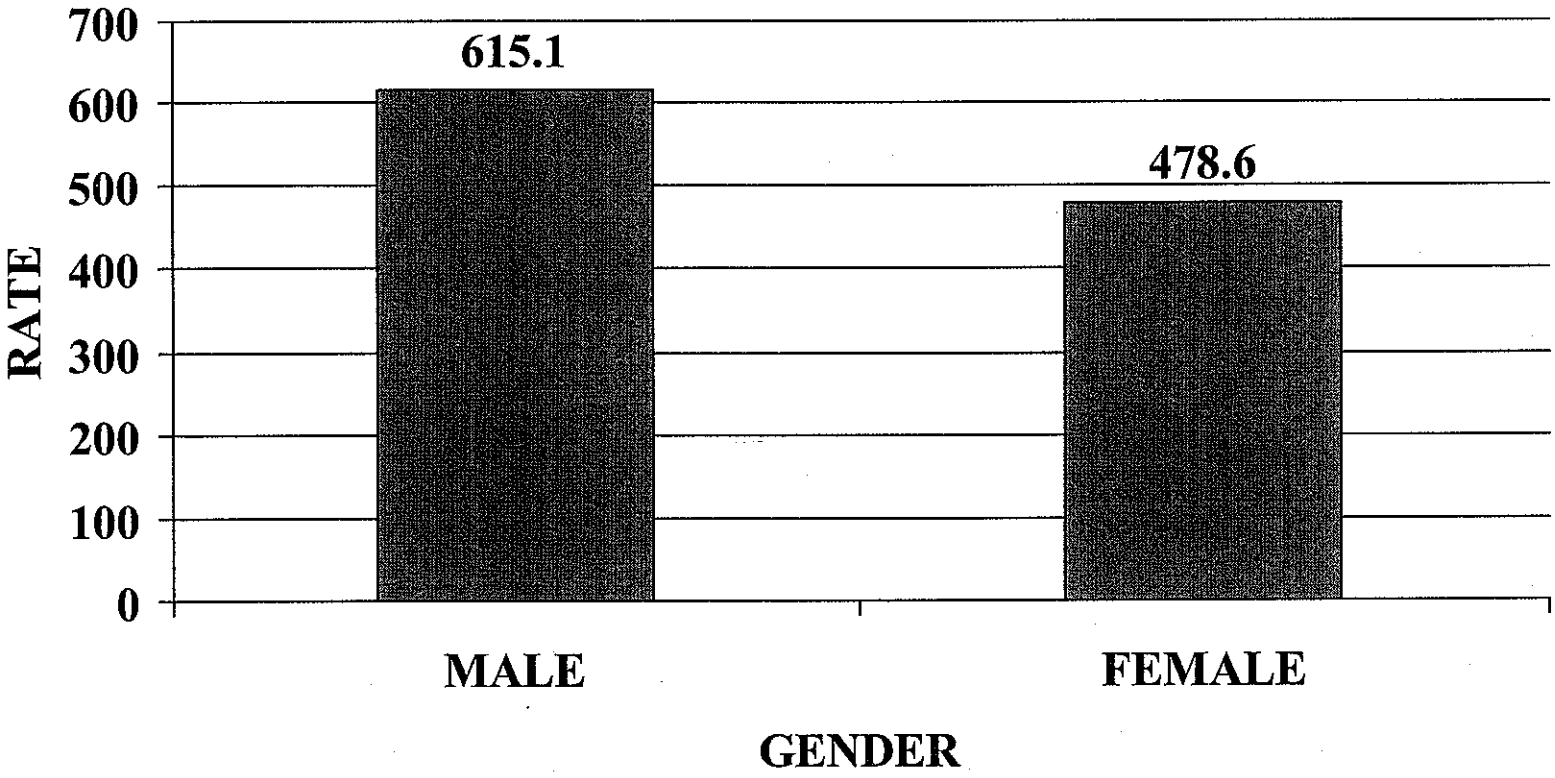
DIAGNOSIS	MALE 172,222		FEMALE 161,990	
	N	Rate	N	Rate
Choanal atresia/stenosis	29	1.7	21	1.3
Lung cysts	23	1.3	5	0.3
Hypoplastic lung	112	6.5	97	6.0
OROFACIAL & GASTROINTESTINAL	**1207**	**70.1**	**861**	**53.2**
Oral clefts	377	21.9	294	18.1
Cleft palate (isolated)	111	6.4	135	8.3
Cleft lip (±cleft palate)	266	15.4	160	9.9
TE fistula/Esophageal atresia	38	2.2	33	2.0
Pyloric stenosis	223	12.9	48	3.0
Atresia/stenosis small intestine	53	3.1	42	2.6
Anal atresia/stenosis	98	5.7	71	4.4
Hirschsprung's disease	59	3.4	25	1.5
Malrotation	52	3.0	45	2.8
Biliary atresia	18	1.0	18	1.1
GENITAL & URINARY	**3227**	**187.4**	**545**	**33.6**
Ovarian cyst	0	0.0	18	1.1
Undescended testicle*	1138	66.1	0	0.0
Hypospadias/Epispadias/Chordee	1136	66.0	0	0.0
Ambiguous genitalia	5	0.3	9	0.6
Absent/hypoplastic kidney	93	5.4	73	4.5
Cystic kidney	78	4.5	74	4.6
Hydronephrosis	309	17.9	124	7.7
Exstrophy bladder	4	0.2	5	0.3
Posterior urethral valves	19	1.1	1	0.1
LIMB & MUSCULOSKELETAL	**2375**	**137.9**	**2234**	**137.9**
Scoliosis	43	2.5	43	2.7
Hip dislocation	98	5.7	262	16.2
Clubfoot	603	35.0	522	32.2
Polydactyly	347	20.1	251	15.5
Syndactyly	173	10.0	110	6.8
Limb reduction deformity - upper	78	4.5	41	2.5
Limb reduction deformity - lower	29	1.7	21	1.3
Craniosynostosis	97	5.6	78	4.8
Crouzon's disease	0	0.0	2	0.1
Apert syndrome	4	0.2	7	0.4
Goldenhar syndrome	11	0.6	8	0.5
Spina bifida occulta	7	0.4	5	0.3
Dwarfism	46	2.7	50	3.1
Osteogenesis imperfecta	6	0.3	9	0.6

TABLE 4. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY GENDER, HAWAII, 1986-2003

DIAGNOSIS	MALE 172,222		FEMALE 161,990	
	N	Rate	N	Rate
Diaphragmatic hernia	46	2.7	37	2.3
Abdominal wall defects	164	9.5	126	7.8
Omphalocele	50	2.9	39	2.4
Gastroschisis	66	3.8	50	3.1
Prune-belly syndrome	7	0.4	2	0.1
Poland syndrome	5	0.3	0	0.0
SKIN & INTEGUMENT	**1254**	**72.8**	**901**	**55.6**
CHROMOSOME & SYNDROME	**928**	**53.9**	**967**	**59.7**
Down syndrome (trisomy 21)	271	15.7	244	15.1
Patau syndrome (trisomy 13)	35	2.0	30	1.9
Edwards syndrome (trisomy 18)	79	4.6	80	4.9
Cri du Chat syndrome	3	0.2	5	0.3
Wolff-Hirschorn syndrome	3	0.2	5	0.3
Autosomal anomalies in normal infant	62	3.6	75	4.6
Triploidy	24	1.4	20	1.2
Turner syndrome (45,X)	1	0.1	139	8.6
Klinefelter syndrome (47,XXY)	51	3.0	0	0.0
Absent spleen/Asplenia	3	0.2	8	0.5
Accessory spleen	26	1.5	15	0.9
Situs inversus	19	1.1	18	1.1
Conjoined twins	4	0.2	0	0.0
Tuberous sclerosis	7	0.4	6	0.4
Amniotic band syndrome	40	2.3	32	2.0
DiGeorge syndrome	12	0.7	9	0.6
Pierre-Robin sequence	17	1.0	14	0.9
Werdnig-Hoffman syndrome	8	0.5	5	0.3
Wolff-Parkinson-White syndrome	10	0.6	8	0.5
NEOPLASMS	**145**	**8.4**	**201**	**12.4**
CONGENITAL INFECTION	**145**	**8.4**	**119**	**7.3**
MATERNAL SUBSTANCE ABUSE	**1455**	**84.5**	**1363**	**84.1**
Fetal alcohol syndrome	51	3.0	36	2.2
Illicit drugs	1424	82.7	1343	82.9
Marijuana	460	26.7	444	27.4
Cocaine	293	17.0	269	16.6
Methamphetamine/Amphetamine	922	53.5	875	54.0
TOTAL	**10594**	**615.1**	**7753**	**478.6**

*birth weight >2499 grams.

FIGURE 4. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY GENDER, HAWAII, 1986-2003



BIRTH DEFECTS BY MATERNAL ETHNICITY

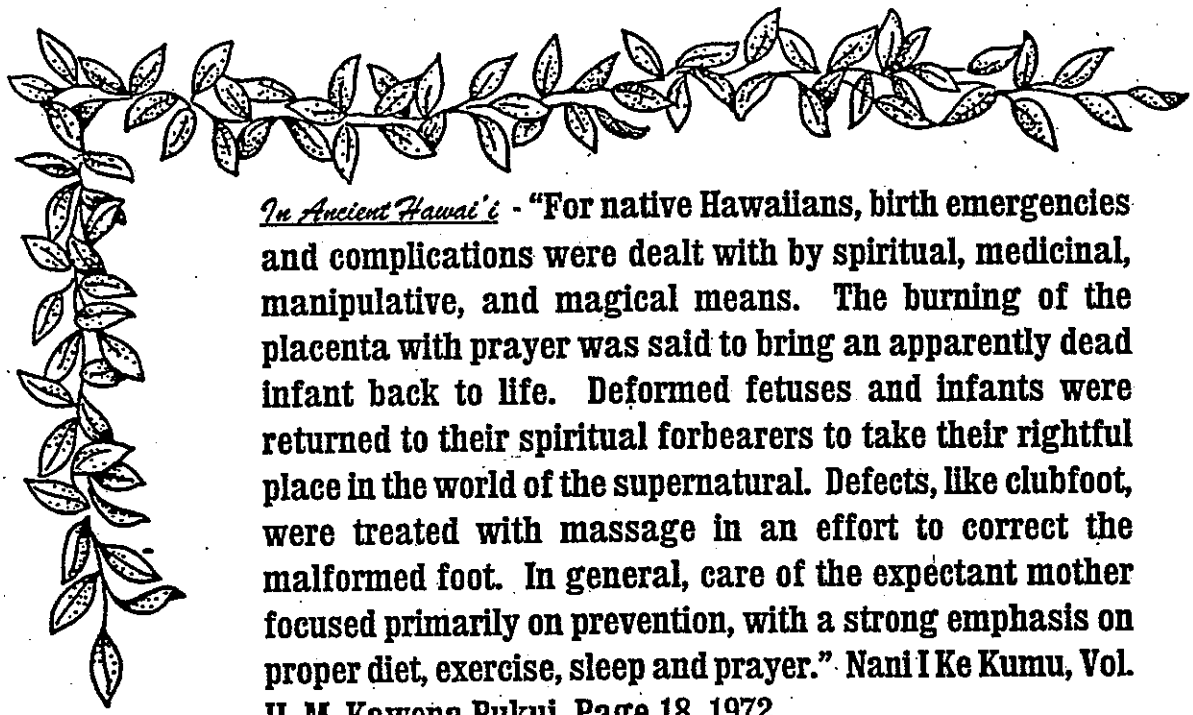
A large proportion of Hawaii's population is of mixed ancestry. A single ethnicity has been assigned to each mother according to guidelines modified from those provided by the CDC and State of Hawaii Department Of Health and outlined as follows:

1) If the woman is noted to be of a single ethnicity, that ethnicity is listed.

2) If the woman is noted to be of more than one ethnicity and one is Caucasian and the other is not, the non-Caucasian ethnicity is listed.

3) If the woman is noted to be of more than one ethnicity and one is Hawaiian or part-Hawaiian, the ethnicity is listed as Hawaiian.

4) If the woman is noted to be of more than one ethnicity and none are Caucasian or Hawaiian, then the first ethnicity noted is listed.



In Ancient Hawai'i - **"For native Hawaiians, birth emergencies and complications were dealt with by spiritual, medicinal, manipulative, and magical means. The burning of the placenta with prayer was said to bring an apparently dead infant back to life. Deformed fetuses and infants were returned to their spiritual forbearers to take their rightful place in the world of the supernatural. Defects, like clubfoot, were treated with massage in an effort to correct the malformed foot. In general, care of the expectant mother focused primarily on prevention, with a strong emphasis on proper diet, exercise, sleep and prayer." Nani I Ke Kumu, Vol. II, M. Kawena Pukui, Page 18, 1972.**

TABLE 5. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY MATERNAL ETHNICITY, HAWAII, 1986-2003

DIAGNOSIS	CAUCASIAN		HAWAIIAN		FILIPINO		JAPANESE		CHINESE		BLACK		SAMOAN	
	82,197		84,219		62,143		41,364		12,432		10,842		9,448	
	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
BRAIN & NERVOUS SYSTEM	**326**	**39.7**	**378**	**44.9**	**255**	**41.0**	**145**	**35.1**	**42**	**33.8**	**39**	**36.0**	**53**	**56.1**
Neural tube defects	93	11.3	77	9.1	48	7.7	43	10.4	11	8.8	6	5.5	14	14.8
Anencephaly	32	3.9	30	3.6	14	2.3	17	4.1	5	4.0	1	0.9	3	3.2
Spina bifida	44	5.4	36	4.3	21	3.4	15	3.6	5	4.0	3	2.8	8	8.5
Encephalocele	17	2.1	14	1.7	14	2.3	11	2.7	2	1.6	3	2.8	3	3.2
Microcephaly	70	8.5	102	12.1	83	13.4	27	6.5	7	5.6	10	9.2	7	7.4
Holoprosencephaly	4	0.5	8	0.9	10	1.6	5	1.2	3	2.4	1	0.9	0	0.0
Hydrocephaly (w/o spina bifida)	88	10.7	106	12.6	68	10.9	34	8.2	9	7.2	14	12.9	14	14.8
EYE, EAR, FACE, & NECK	**407**	**49.5**	**448**	**53.2**	**354**	**57.0**	**190**	**45.9**	**67**	**53.9**	**47**	**43.3**	**55**	**58.2**
EYE	173	21.0	174	20.7	130	20.9	72	17.4	24	19.3	9	8.3	17	18.0
Anophthalmia	1	0.1	5	0.6	2	0.3	1	0.2	0	0.0	0	0.0	1	1.1
Microphthalmia	31	3.8	27	3.2	11	1.8	12	2.9	6	4.8	1	0.9	3	3.2
Cataract	14	1.7	12	1.4	4	0.6	4	1.0	1	0.8	3	2.8	0	0.0
EAR	284	34.6	316	37.5	265	42.6	133	32.2	48	38.6	34	31.4	40	42.3
Absent auditory canal/ear	4	0.5	10	1.2	9	1.4	4	1.0	1	0.8	0	0.0	3	3.2
Microtia	15	1.8	34	4.0	26	4.2	13	3.1	6	4.8	2	1.8	8	8.5
CARDIAC & CIRCULATORY	**1362**	**165.7**	**1627**	**193.2**	**1295**	**208.4**	**683**	**165.1**	**206**	**165.7**	**202**	**186.3**	**254**	**268.8**
Common truncus	6	0.7	3	0.4	4	0.6	5	1.2	1	0.8	3	2.8	1	1.1
Transposition of the great vessels	37	4.5	48	5.7	21	3.4	12	2.9	6	4.8	0	0.0	2	2.1
Tetralogy of Fallot	18	2.2	44	5.2	28	4.5	20	4.8	3	2.4	4	3.7	1	1.1
Single ventricle	7	0.9	8	0.9	4	0.6	3	0.7	0	0.0	1	0.9	0	0.0
Ventricular septal defect	400	48.7	394	46.8	299	48.1	204	49.3	60	48.3	45	41.5	52	55.0
Atrial septal defect	424	51.6	640	76.0	542	87.2	236	57.1	69	55.5	55	50.7	91	96.3
Endocardial cushion defect	24	2.9	22	2.6	11	1.8	8	1.9	5	4.0	2	1.8	1	1.1
Pulmonary valve atresia/stenosis	58	7.1	94	11.2	60	9.7	53	12.8	10	8.0	10	9.2	7	7.4
Tricuspid valve atresia/stenosis	10	1.2	25	3.0	6	1.0	5	1.2	3	2.4	1	0.9	1	1.1
Ebstein's anomaly	6	0.7	2	0.2	2	0.3	4	1.0	0	0.0	0	0.0	1	1.1

TABLE 5. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY MATERNAL ETHNICITY, HAWAII, 1986-2003

DIAGNOSIS	KOREAN		PORTUGUESE		VIETNAMESE		OTHER	
	6,824		3,006		2,568		17,133	
	N	Rate	N	Rate	N	Rate	N	Rate
BRAIN & NERVOUS SYSTEM	**24**	**35.2**	**20**	**66.5**	**8**	**31.2**	**103**	**60.1**
Neural tube defects	4	5.9	4	13.3	2	7.8	19	11.1
Anencephaly	4	5.9	1	3.3	1	3.9	5	2.9
Spina bifida	0	0.0	3	10.0	1	3.9	12	7.0
Encephalocele	0	0.0	1	3.3	0	0.0	3	1.8
Microcephaly	8	11.7	5	16.6	0	0.0	25	14.6
Holoprosencephaly	2	2.9	0	0.0	1	3.9	6	3.5
Hydrocephaly (w/o spina bifida)	6	8.8	4	13.3	1	3.9	29	16.9
EYE, EAR, FACE, & NECK	**36**	**52.8**	**20**	**66.5**	**8**	**31.2**	**153**	**89.3**
EYE	14	20.5	9	29.9	3	11.7	61	35.6
Anophthalmia	0	0.0	0	0.0	0	0.0	3	1.8
Microphthalmia	2	2.9	0	0.0	0	0.0	11	6.4
Cataract	1	1.5	0	0.0	0	0.0	5	2.9
EAR	24	35.2	14	46.6	4	15.6	97	56.6
Absent auditory canal/ear	2	2.9	0	0.0	0	0.0	4	2.3
Microtia	1	1.5	2	6.7	0	0.0	13	7.6
CARDIAC & CIRCULATORY	**98**	**143.6**	**72**	**239.5**	**49**	**190.8**	**557**	**325.1**
Common truncus	0	0.0	0	0.0	1	3.9	0	0.0
Transposition of the great vessels	3	4.4	1	3.3	1	3.9	13	7.6
Tetralogy of Fallot	4	5.9	1	3.3	1	3.9	7	4.1
Single ventricle	2	2.9	0	0.0	0	0.0	3	1.8
Ventricular septal defect	27	39.6	14	46.6	12	46.7	106	61.9
Atrial septal defect	41	60.1	23	76.5	22	85.7	206	120.2
Endocardial cushion defect	0	0.0	0	0.0	0	0.0	7	4.1
Pulmonary valve atresia/stenosis	6	8.8	1	3.3	3	11.7	28	16.3
Tricuspid valve atresia/stenosis	0	0.0	0	0.0	1	3.9	4	2.3
Ebstein's anomaly	0	0.0	0	0.0	0	0.0	2	1.2

TABLE 5. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY MATERNAL ETHNICITY, HAWAII, 1986-2003

DIAGNOSIS	CAUCASIAN 82,197		HAWAIIAN 84,219		FILIPINO 62,143		JAPANESE 41,364		CHINESE 12,432		BLACK 10,842		SAMOAN 9,448	
	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Aortic valve atresia/stenosis	12	1.5	7	0.8	7	1.1	10	2.4	3	2.4	2	1.8	3	3.2
Mitral valve atresia/stenosis	6	0.7	12	1.4	5	0.8	7	1.7	1	0.8	0	0.0	2	2.1
Hypoplastic left heart syndrome	14	1.7	13	1.5	7	1.1	6	1.5	2	1.6	1	0.9	2	2.1
Dextrocardia	17	2.1	15	1.8	2	0.3	5	1.2	3	2.4	3	2.8	3	3.2
Patent ductus arteriosus*	503	61.2	678	80.5	570	91.7	260	62.9	99	79.6	66	60.9	138	146.1
Coarctation of the aorta	37	4.5	17	2.0	20	3.2	9	2.2	0	0.0	2	1.8	1	1.1
Aorta stenosis	0	0.0	0	0.0	1	0.2	0	0.0	0	0.0	0	0.0	1	1.1
Pulmonary artery atresia/stenosis	113	13.7	172	20.4	137	22.0	57	13.8	19	15.3	32	29.5	30	31.8
Total anomalous pulmonary venous return	4	0.5	5	0.6	7	1.1	8	1.9	3	2.4	1	0.9	1	1.1
Partial anomalous pulmonary venous	1	0.1	4	0.5	3	0.5	3	0.7	0	0.0	2	1.8	1	1.1
Persistent fetal circulation	48	5.8	44	5.2	45	7.2	18	4.4	9	7.2	4	3.7	15	15.9
RESPIRATORY	**234**	**28.5**	**217**	**25.8**	**175**	**28.2**	**88**	**21.3**	**30**	**24.1**	**39**	**36.0**	**26**	**27.5**
Choanal atresia/stenosis	14	1.7	9	1.1	13	2.1	4	1.0	3	2.4	3	2.8	0	0.0
Lung cysts	3	0.4	13	1.5	7	1.1	2	0.5	0	0.0	0	0.0	2	2.1
Hypoplastic lung	55	6.7	46	5.5	36	5.8	28	6.8	5	4.0	10	9.2	6	6.4
OROFACIAL & GASTROINTESTINAL	**545**	**66.3**	**466**	**55.3**	**350**	**56.3**	**263**	**63.6**	**83**	**66.8**	**66**	**60.9**	**69**	**73.0**
Oral clefts	129	15.7	168	19.9	128	20.6	104	25.1	36	29.0	16	14.8	21	22.2
Cleft palate (isolated)	49	6.0	78	9.3	38	6.1	35	8.5	15	12.1	3	2.8	5	5.3
Cleft lip (±cleft palate)	80	9.7	90	10.7	90	14.5	69	16.7	22	17.7	13	12.0	16	16.9
TE fistula/Esophageal atresia	24	2.9	15	1.8	10	1.6	7	1.7	2	1.6	3	2.8	1	1.1
Pyloric stenosis	137	16.7	44	5.2	13	2.1	34	8.2	5	4.0	12	11.1	0	0.0
Atresia/stenosis small intestine	21	2.6	25	3.0	13	2.1	20	4.8	3	2.4	1	0.9	2	2.1
Anal atresia/stenosis	39	4.7	43	5.1	32	5.1	21	5.1	9	7.2	5	4.6	5	5.3
Hirschsprung's disease	16	1.9	11	1.3	19	3.1	9	2.2	7	5.6	6	5.5	5	5.3
Malrotation	18	2.2	26	3.1	17	2.7	12	2.9	4	3.2	3	2.8	6	6.4
Biliary atresia	6	0.7	8	0.9	8	1.3	4	1.0	1	0.8	0	0.0	2	2.1
GENITAL & URINARY	**955**	**116.2**	**931**	**110.5**	**752**	**121.0**	**344**	**83.2**	**98**	**78.8**	**157**	**144.8**	**126**	**133.4**

TABLE 5. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY MATERNAL ETHNICITY, HAWAII, 1986-2003

DIAGNOSIS	KOREAN		PORTUGUESE		VIETNAMESE		OTHER	
	6,824		3,006		2,568		17,133	
	N	Rate	N	Rate	N	Rate	N	Rate
Aortic valve atresia/stenosis	2	2.9	2	6.7	0	0.0	2	1.2
Mitral valve atresia/stenosis	0	0.0	1	3.3	0	0.0	3	1.8
Hypoplastic left heart syndrome	1	1.5	1	3.3	0	0.0	3	1.8
Dextrocardia	2	2.9	0	0.0	0	0.0	3	1.8
Patent ductus arteriosus*	41	60.1	30	99.8	24	93.5	280	163.4
Coarctation of the aorta	2	2.9	1	3.3	1	3.9	7	4.1
Aorta stenosis	0	0.0	0	0.0	0	0.0	0	0.0
Pulmonary artery atresia/stenosis	5	7.3	4	13.3	8	31.2	60	35.0
Total anomalous pulmonary venous return	1	1.5	0	0.0	0	0.0	2	1.2
Partial anomalous pulmonary venous	0	0.0	0	0.0	0	0.0	3	1.8
Persistent fetal circulation	1	1.5	1	3.3	1	3.9	14	8.2
RESPIRATORY	**22**	**32.2**	**17**	**56.6**	**3**	**11.7**	**54**	**31.5**
Choanal atresia/stenosis	2	2.9	1	3.3	0	0.0	1	0.6
Lung cysts	0	0.0	0	0.0	0	0.0	1	0.6
Hypoplastic lung	4	5.9	2	6.7	0	0.0	16	9.3
OROFACIAL & GASTROINTESTINAL	**47**	**68.9**	**26**	**86.5**	**14**	**54.5**	**139**	**81.1**
Oral clefts	17	24.9	10	33.3	5	19.5	42	24.5
Cleft palate (isolated)	6	8.8	3	10.0	1	3.9	14	8.2
Cleft lip (±cleft palate)	11	16.1	7	23.3	4	15.6	28	16.3
TE fistula/Esophageal atresia	2	2.9	0	0.0	0	0.0	7	4.1
Pyloric stenosis	7	10.3	4	13.3	1	3.9	15	8.8
Atresia/stenosis small intestine	2	2.9	0	0.0	1	3.9	7	4.1
Anal atresia/stenosis	2	2.9	1	3.3	1	3.9	9	5.3
Hirschsprung's disease	2	2.9	1	3.3	0	0.0	7	4.1
Malrotation	0	0.0	1	3.3	0	0.0	9	5.3
Biliary atresia	0	0.0	2	6.7	0	0.0	5	2.9
GENITAL & URINARY	**66**	**96.7**	**38**	**126.4**	**18**	**70.1**	**279**	**162.8**

TABLE 5. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY MATERNAL ETHNICITY, HAWAII, 1986-2003

DIAGNOSIS	CAUCASIAN		HAWAIIAN		FILIPINO		JAPANESE		CHINESE		BLACK		SAMOAN	
	82,197		84,219		62,143		41,364		12,432		10,842		9,448	
	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Ovarian cyst	7	0.9	2	0.2	3	0.5	4	1.0	0	0.0	1	0.9	0	0.0
Undescended testicle*	270	32.8	324	38.5	244	39.3	91	22.0	19	15.3	52	48.0	31	32.8
Hypospadias/Epispadias/Chordee	308	37.5	256	30.4	242	38.9	99	23.9	30	24.1	40	36.9	37	39.2
Ambiguous genitalia	4	0.5	4	0.5	4	0.6	1	0.2	1	0.8	0	0.0	0	0.0
Absent/hypoplastic kidney	45	5.5	37	4.4	37	6.0	12	2.9	8	6.4	7	6.5	5	5.3
Cystic kidney	40	4.9	32	3.8	23	3.7	17	4.1	4	3.2	7	6.5	9	9.5
Hydronephrosis	127	15.5	93	11.0	70	11.3	44	10.6	14	11.3	16	14.8	18	19.1
Exstrophy bladder	2	0.2	1	0.1	2	0.3	1	0.2	0	0.0	0	0.0	0	0.0
Posterior urethral valves	8	1.0	5	0.6	1	0.2	2	0.5	0	0.0	3	2.8	1	1.1
LIMB & MUSCULOSKELETAL	**1274**	**155.0**	**1072**	**127.3**	**740**	**119.1**	**512**	**123.8**	**155**	**124.7**	**206**	**190.0**	**147**	**155.6**
Scoliosis	20	2.4	22	2.6	15	2.4	12	2.9	1	0.8	1	0.9	2	2.1
Hip dislocation	118	14.4	63	7.5	60	9.7	69	16.7	18	14.5	1	0.9	5	5.3
Clubfoot	322	39.2	325	38.6	154	24.8	90	21.8	29	23.3	39	36.0	45	47.6
Polydactyly	108	13.1	131	15.6	70	11.3	56	13.5	27	21.7	101	93.2	26	27.5
Syndactyly	99	12.0	73	8.7	33	5.3	25	6.0	8	6.4	6	5.5	8	8.5
Limb reduction deformity - upper	22	2.7	38	4.5	25	4.0	10	2.4	4	3.2	5	4.6	5	5.3
Limb reduction deformity - lower	7	0.9	16	1.9	10	1.6	7	1.7	1	0.8	2	1.8	3	3.2
Craniosynostosis	69	8.4	31	3.7	26	4.2	25	6.0	6	4.8	4	3.7	0	0.0
Crouzon's disease	0	0.0	1	0.1	1	0.2	0	0.0	0	0.0	0	0.0	0	0.0
Apert syndrome	3	0.4	2	0.2	3	0.5	1	0.2	1	0.8	0	0.0	0	0.0
Goldenhar syndrome	2	0.2	6	0.7	3	0.5	3	0.7	1	0.8	1	0.9	1	1.1
Spina bifida occulta	6	0.7	2	0.2	2	0.3	2	0.5	0	0.0	0	0.0	0	0.0
Dwarfism	25	3.0	17	2.0	19	3.1	19	4.6	1	0.8	0	0.0	1	1.1
Osteogenesis imperfecta	5	0.6	4	0.5	2	0.3	1	0.2	0	0.0	0	0.0	1	1.1
Diaphragmatic hernia	28	3.4	17	2.0	16	2.6	10	2.4	1	0.8	1	0.9	4	4.2
Abdominal wall defects	85	10.3	67	8.0	57	9.2	27	6.5	6	4.8	10	9.2	5	5.3
Omphalocele	26	3.2	15	1.8	20	3.2	8	1.9	5	4.0	3	2.8	1	1.1

TABLE 5. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY MATERNAL ETHNICITY, HAWAII, 1986-2003

DIAGNOSIS	KOREAN		PORTUGUESE		VIETNAMESE		OTHER	
	6,824		3,006		2,568		17,133	
	N	Rate	N	Rate	N	Rate	N	Rate
Ovarian cyst	0	0.0	0	0.0	0	0.0	1	0.6
Undescended testicle*	18	26.4	16	53.2	7	27.3	65	37.9
Hypospadias/Epispadias/Chordee	26	38.1	10	33.3	5	19.5	81	47.3
Ambiguous genitalia	0	0.0	0	0.0	0	0.0	1	0.6
Absent/hypoplastic kidney	1	1.5	0	0.0	0	0.0	10	5.8
Cystic kidney	2	2.9	0	0.0	0	0.0	16	9.3
Hydronephrosis	5	7.3	6	20.0	1	3.9	36	21.0
Exstrophy bladder	1	1.5	0	0.0	1	3.9	1	0.6
Posterior urethral valves	1	1.5	0	0.0	0	0.0	0	0.0
LIMB & MUSCULOSKELETAL	**80**	**117.2**	**46**	**153.0**	**32**	**124.6**	**343**	**200.2**
Scoliosis	1	1.5	1	3.3	0	0.0	8	4.7
Hip dislocation	7	10.3	1	3.3	1	3.9	17	9.9
Clubfoot	13	19.1	15	49.9	3	11.7	87	50.8
Polydactyly	11	16.1	3	10.0	7	27.3	59	34.4
Syndactyly	1	1.5	1	3.3	3	11.7	27	15.8
Limb reduction deformity - upper	2	2.9	1	3.3	1	3.9	6	3.5
Limb reduction deformity - lower	2	2.9	1	3.3	1	3.9	0	0.0
Craniosynostosis	2	2.9	2	6.7	0	0.0	10	5.8
Crouzon's disease	0	0.0	0	0.0	0	0.0	0	0.0
Apert syndrome	0	0.0	0	0.0	0	0.0	1	0.6
Goldenhar syndrome	0	0.0	0	0.0	0	0.0	2	1.2
Spina bifida occulta	0	0.0	0	0.0	0	0.0	0	0.0
Dwarfism	5	7.3	1	3.3	0	0.0	7	4.1
Osteogenesis imperfecta	0	0.0	0	0.0	1	3.9	1	0.6
Diaphragmatic hernia	1	1.5	0	0.0	2	7.8	3	1.8
Abdominal wall defects	9	13.2	4	13.3	2	7.8	20	11.7
Omphalocele	6	8.8	0	0.0	1	3.9	6	3.5

TABLE 5. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY MATERNAL ETHNICITY, HAWAII, 1986-2003

DIAGNOSIS	CAUCASIAN 82,197		HAWAIIAN 84,219		FILIPINO 62,143		JAPANESE 41,364		CHINESE 12,432		BLACK 10,842		SAMOAN 9,448	
	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Gastroschisis	27	3.3	35	4.2	20	3.2	12	2.9	1	0.8	4	3.7	4	4.2
Prune-belly syndrome	4	0.5	1	0.1	3	0.5	0	0.0	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	0	0.0
SKIN & INTEGUMENT	**300**	**36.5**	**552**	**65.5**	**468**	**75.3**	**273**	**66.0**	**86**	**69.2**	**110**	**101.5**	**90**	**95.3**
CHROMOSOME & SYNDROME	**514**	**62.5**	**300**	**35.6**	**314**	**50.5**	**318**	**76.9**	**103**	**82.9**	**42**	**38.7**	**41**	**43.4**
Down syndrome (trisomy 21)	141	17.2	80	9.5	93	15.0	81	19.6	35	28.2	4	3.7	8	8.5
Patau syndrome (trisomy 13)	22	2.7	8	0.9	8	1.3	8	1.9	7	5.6	2	1.8	1	1.1
Edwards syndrome (trisomy 18)	46	5.6	19	2.3	25	4.0	29	7.0	9	7.2	3	2.8	2	2.1
Cri du Chat syndrome	3	0.4	0	0.0	1	0.2	0	0.0	1	0.8	0	0.0	2	2.1
Wolff-Hirschorn syndrome	1	0.1	0	0.0	2	0.3	3	0.7	0	0.0	0	0.0	0	0.0
Autosomal anomalies in normal infant	35	4.3	18	2.1	29	4.7	30	7.3	11	8.8	3	2.8	1	1.1
Triploidy	13	1.6	6	0.7	3	0.5	6	1.5	3	2.4	1	0.9	0	0.0
Turner syndrome (45,X)	36	4.4	18	2.1	20	3.2	30	7.3	6	4.8	6	5.5	1	1.1
Klinefelter syndrome (47,XXY)	10	1.2	6	0.7	9	1.4	15	3.6	2	1.6	1	0.9	0	0.0
Absent spleen/Asplenia	1	0.1	5	0.6	1	0.2	1	0.2	1	0.8	0	0.0	0	0.0
Accessory spleen	9	1.1	10	1.2	9	1.4	6	1.5	1	0.8	0	0.0	4	4.2
Situs inversus	6	0.7	12	1.4	6	1.0	5	1.2	1	0.8	0	0.0	1	1.1
Conjoined twins	2	0.2	2	0.2	2	0.3	0	0.0	0	0.0	0	0.0	2	2.1
Tuberous sclerosis	3	0.4	4	0.5	2	0.3	1	0.2	1	0.8	1	0.9	0	0.0
Amniotic band syndrome	18	2.2	25	3.0	11	1.8	7	1.7	1	0.8	4	3.7	0	0.0
DiGeorge syndrome	5	0.6	7	0.8	1	0.2	4	1.0	0	0.0	1	0.9	0	0.0
Pierre-Robin sequence	7	0.9	8	0.9	6	1.0	6	1.5	2	1.6	1	0.9	0	0.0
Werdnig-Hoffman syndrome	3	0.4	3	0.4	0	0.0	0	0.0	0	0.0	1	0.9	3	3.2
Wolff-Parkinson-White syndrome	8	1.0	5	0.6	3	0.5	1	0.2	0	0.0	0	0.0	1	1.1
NEOPLASMS	**80**	**9.7**	**74**	**8.8**	**66**	**10.6**	**52**	**12.6**	**14**	**11.3**	**12**	**11.1**	**10**	**10.6**
CONGENITAL INFECTION	**53**	**6.4**	**59**	**7.0**	**60**	**9.7**	**26**	**6.3**	**10**	**8.0**	**13**	**12.0**	**12**	**12.7**
MATERNAL SUBSTANCE ABUSE	**374**	**45.5**	**1553**	**184.4**	**318**	**51.2**	**174**	**42.1**	**26**	**20.9**	**37**	**34.1**	**78**	**82.6**

TABLE 5. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY MATERNAL ETHNICITY, HAWAII, 1986-2003

DIAGNOSIS	KOREAN		PORTUGUESE		VIETNAMESE		OTHER	
	6,824		3,006		2,568		17,133	
	N	Rate	N	Rate	N	Rate	N	Rate
Gastroschisis	3	4.4	1	3.3	1	3.9	8	4.7
Prune-belly syndrome	0	0.0	0	0.0	0	0.0	1	0.6
Poland syndrome	0	0.0	0	0.0	0	0.0	0	0.0
SKIN & INTEGUMENT	**47**	**68.9**	**21**	**69.9**	**20**	**77.9**	**181**	**105.6**
CHROMOSOME & SYNDROME	**40**	**58.6**	**19**	**63.2**	**7**	**27.3**	**112**	**65.4**
Down syndrome (trisomy 21)	7	10.3	6	20.0	2	7.8	30	17.5
Patau syndrome (trisomy 13)	2	2.9	1	3.3	0	0.0	4	2.3
Edwards syndrome (trisomy 18)	3	4.4	1	3.3	1	3.9	8	4.7
Cri du Chat syndrome	0	0.0	0	0.0	0	0.0	0	0.0
Wolff-Hirschorn syndrome	1	1.5	0	0.0	0	0.0	0	0.0
Autosomal anomalies in normal infant	5	7.3	0	0.0	0	0.0	4	2.3
Triploidy	0	0.0	1	3.3	0	0.0	0	0.0
Turner syndrome (45,X)	2	2.9	2	6.7	1	3.9	8	4.7
Klinefelter syndrome (47,XXY)	2	2.9	0	0.0	0	0.0	5	2.9
Absent spleen/Asplenia	1	1.5	0	0.0	0	0.0	1	0.6
Accessory spleen	0	0.0	0	0.0	0	0.0	2	1.2
Situs inversus	2	2.9	0	0.0	0	0.0	3	1.8
Conjoined twins	2	2.9	2	6.7	0	0.0	0	0.0
Tuberous sclerosis	1	1.5	0	0.0	0	0.0	0	0.0
Amniotic band syndrome	2	2.9	1	3.3	1	3.9	3	1.8
DiGeorge syndrome	2	2.9	0	0.0	0	0.0	1	0.6
Pierre-Robin sequence	0	0.0	1	3.3	0	0.0	0	0.0
Werdnig-Hoffman syndrome	0	0.0	0	0.0	0	0.0	3	1.8
Wolff-Parkinson-White syndrome	0	0.0	0	0.0	0	0.0	1	0.6
NEOPLASMS	**9**	**13.2**	**6**	**20.0**	**0**	**0.0**	**20**	**11.7**
CONGENITAL INFECTION	**1**	**1.5**	**2**	**6.7**	**1**	**3.9**	**27**	**15.8**
MATERNAL SUBSTANCE ABUSE	**43**	**63.0**	**47**	**156.4**	**4**	**15.6**	**181**	**105.6**

TABLE 5. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY MATERNAL ETHNICITY, HAWAII, 1986-2003

DIAGNOSIS	CAUCASIAN		HAWAIIAN		FILIPINO		JAPANESE		CHINESE		BLACK		SAMOAN	
	82,197		84,219		62,143		41,364		12,432		10,842		9,448	
	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Fetal alcohol syndrome	26	3.2	42	5.0	5	0.8	4	1.0	1	0.8	1	0.9	0	0.0
Illicit drugs	358	43.6	1529	181.6	314	50.5	171	41.3	26	20.9	37	34.1	78	82.6
Marijuana	148	18.0	507	60.2	75	12.1	38	9.2	8	6.4	18	16.6	28	29.6
Cocaine	117	14.2	277	32.9	26	4.2	34	8.2	7	5.6	15	13.8	10	10.6
Methamphetamine/Amphetamine	140	17.0	1057	125.5	245	39.4	126	30.5	14	11.3	12	11.1	51	54.0
TOTAL	**4365**	**531.0**	**4997**	**593.3**	**3146**	**506.3**	**1987**	**480.4**	**569**	**457.7**	**642**	**592.1**	**606**	**641.4**

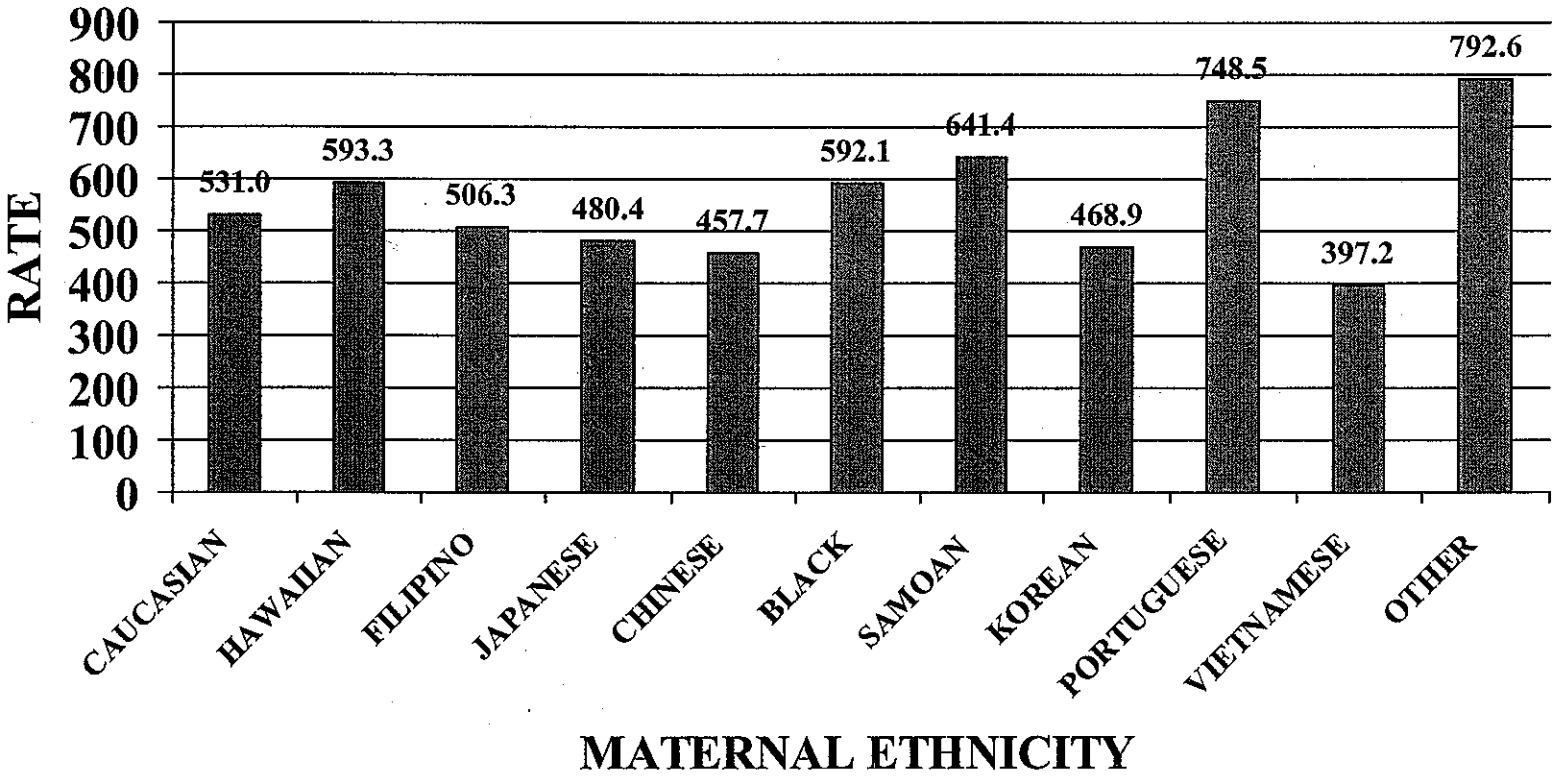
TABLE 5. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY MATERNAL ETHNICITY, HAWAII, 1986-2003

DIAGNOSIS	KOREAN		PORTUGUESE		VIETNAMESE		OTHER	
	6,824		3,006		2,568		17,133	
	N	Rate	N	Rate	N	Rate	N	Rate
Fetal alcohol syndrome	1	1.5	0	0.0	2	7.8	7	4.1
Illicit drugs	42	61.5	47	156.4	4	15.6	178	103.9
Marijuana	10	14.7	18	59.9	1	3.9	62	36.2
Cocaine	8	11.7	21	69.9	3	11.7	51	29.8
Methamphetamine/Amphetamine	31	45.4	23	76.5	1	3.9	101	59.0
TOTAL	**320**	**468.9**	**225**	**748.5**	**102**	**397.2**	**1358**	**792.6**

*birth weight >2499 grams.

In previous databooks, individual analyses were performed for Hispanics, Native Americans, and Guamanians. Denominators for these groups were not available for 2003, so these groups were included in the "other" group.

FIGURE 5. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY MATERNAL ETHNICITY, HAWAII, 1986-2003



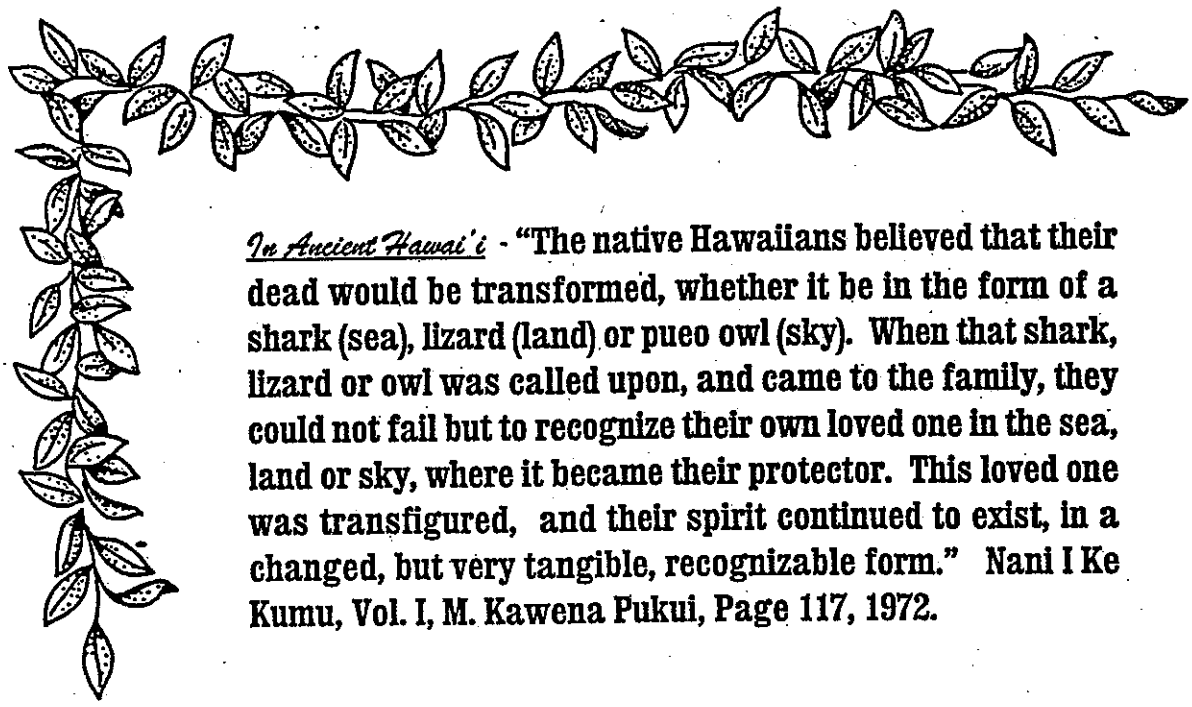
RESIDENCE AT BIRTH AND CONCEPTION

1) The addresses for 60 (0.3%) of the cases in the HBDP data base delivered between 1986 and 2003 are not known.

2) Of the 18,495 cases where the residence at delivery is known, 202 (1.1%) lived outside of Hawaii.

3) Of the 18,293 cases who resided in Hawaii at delivery, 1,245 (6.8%) are known to have lived at a different address at the time of conception, 575 (46.2%) conceived outside of Hawaii and 670 (53.8%) conceived within the state.

4) Thus, of the 18,495 cases with known residence at delivery, 777 (4.2%) are known to have not been conceived in Hawaii.



In Ancient Hawai'i - **"The native Hawaiians believed that their dead would be transformed, whether it be in the form of a shark (sea), lizard (land) or pueo owl (sky). When that shark, lizard or owl was called upon, and came to the family, they could not fail but to recognize their own loved one in the sea, land or sky, where it became their protector. This loved one was transfigured, and their spirit continued to exist, in a changed, but very tangible, recognizable form." Nani I Ke Kumu, Vol. I, M. Kawena Pukui, Page 117, 1972.**

TABLE 6. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY COUNTY, HAWAII, 1986-2003

DIAGNOSIS	HONOLULU		HAWAII		MAUI		KAUAI	
	263,409		38,268		33,380		16,081	
	N	Rate	N	Rate	N	Rate	N	Rate
BRAIN & NERVOUS SYSTEM	**1029**	**39.1**	**176**	**46.0**	**115**	**34.5**	**64**	**39.8**
Neural tube defects	243	9.2	36	9.4	32	9.6	11	6.8
Anencephaly	92	3.5	11	2.9	11	3.3	4	2.5
Spina bifida	108	4.1	14	3.7	18	5.4	5	3.1
Encephalocele	51	1.9	11	2.9	3	0.9	3	1.9
Microcephaly	249	9.5	45	11.8	24	7.2	21	13.1
Holoprosencephaly	27	1.0	5	1.3	7	2.1	2	1.2
Hydrocephaly (w/o spina bifida)	275	10.4	43	11.2	35	10.5	14	8.7
EYE, EAR, FACE, & NECK	**1287**	**48.9**	**222**	**58.0**	**179**	**53.6**	**74**	**46.0**
EYE	463	17.6	109	28.5	68	20.4	29	18.0
Anophthalmia	8	0.3	2	0.5	3	0.9	0	0.0
Microphthalmia	69	2.6	15	3.9	11	3.3	5	3.1
Cataract	29	1.1	11	2.9	2	0.6	0	0.0
EAR	909	34.5	147	38.4	129	38.6	55	34.2
Absent auditory canal/ear	29	1.1	4	1.0	3	0.9	1	0.6
Microtia	93	3.5	11	2.9	11	3.3	5	3.1
CARDIAC & CIRCULATORY	**4940**	**187.5**	**635**	**165.9**	**480**	**143.8**	**291**	**181.0**
Common truncus	20	0.8	0	0.0	1	0.3	2	1.2
Transposition of the great vessels	98	3.7	24	6.3	17	5.1	5	3.1
Tetralogy of Fallot	99	3.8	12	3.1	13	3.9	5	3.1
Single ventricle	19	0.7	3	0.8	5	1.5	1	0.6
Ventricular septal defect	1248	47.4	166	43.4	123	36.8	68	42.3
Atrial septal defect	1751	66.5	254	66.4	213	63.8	107	66.5
Endocardial cushion defect	55	2.1	12	3.1	7	2.1	3	1.9
Pulmonary valve atresia/stenosis	244	9.3	39	10.2	29	8.7	12	7.5
Tricuspid valve atresia/stenosis	46	1.7	6	1.6	1	0.3	1	0.6
Ebstein's anomaly	18	0.7	0	0.0	0	0.0	0	0.0
Aortic valve atresia/stenosis	33	1.3	10	2.6	6	1.8	1	0.6
Mitral valve atresia/stenosis	24	0.9	7	1.8	5	1.5	1	0.6
Hypoplastic left heart syndrome	33	1.3	10	2.6	9	2.7	1	0.6
Dextrocardia	34	1.3	8	2.1	10	3.0	1	0.6
Patent ductus arteriosus*	2162	82.1	205	53.6	210	62.9	108	67.2
Coarctation of the aorta	67	2.5	11	2.9	16	4.8	3	1.9
Aorta stenosis	1	0.0	0	0.0	1	0.3	0	0.0
Pulmonary artery atresia/stenosis	489	18.6	73	19.1	39	11.7	31	19.3
Total anomalous pulmonary venous return	22	0.8	4	1.0	3	0.9	3	1.9
Partial anomalous pulmonary venous	11	0.4	1	0.3	3	0.9	1	0.6
Persistent fetal circulation	139	5.3	19	5.0	27	8.1	16	9.9
RESPIRATORY	**651**	**24.7**	**112**	**29.3**	**89**	**26.7**	**40**	**24.9**

TABLE 6. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY COUNTY, HAWAII, 1986-2003

DIAGNOSIS	HONOLULU		HAWAII		MAUI		KAUAI	
	263,409		38,268		33,380		16,081	
	N	Rate	N	Rate	N	Rate	N	Rate
Choanal atresia/stenosis	36	1.4	7	1.8	6	1.8	1	0.6
Lung cysts	25	0.9	1	0.3	1	0.3	1	0.6
Hypoplastic lung	151	5.7	22	5.7	15	4.5	10	6.2
OROFACIAL & GASTROINTESTINAL	**1533**	**58.2**	**236**	**61.7**	**189**	**56.6**	**88**	**54.7**
Oral clefts	485	18.4	84	22.0	72	21.6	29	18.0
Cleft palate (isolated)	162	6.2	33	8.6	32	9.6	18	11.2
Cleft lip (±cleft palate)	324	12.3	51	13.3	40	12.0	11	6.8
TE fistula/Esophageal atresia	50	1.9	12	3.1	9	2.7	0	0.0
Pyloric stenosis	217	8.2	28	7.3	17	5.1	9	5.6
Atresia/stenosis small intestine	64	2.4	16	4.2	8	2.4	3	1.9
Anal atresia/stenosis	132	5.0	13	3.4	16	4.8	7	4.4
Hirschsprung's disease	68	2.6	10	2.6	3	0.9	2	1.2
Malrotation	76	2.9	8	2.1	6	1.8	3	1.9
Biliary atresia	25	0.9	4	1.0	2	0.6	4	2.5
GENITAL & URINARY	**2832**	**107.5**	**350**	**91.5**	**372**	**111.4**	**172**	**107.0**
Ovarian cyst	13	0.5	2	0.5	2	0.6	1	0.6
Undescended testicle*	850	32.3	107	28.0	126	37.7	50	31.1
Hypospadias/Epispadias/Chordee	835	31.7	108	28.2	127	38.0	56	34.8
Ambiguous genitalia	13	0.5	0	0.0	1	0.3	1	0.6
Absent/hypoplastic kidney	120	4.6	18	4.7	15	4.5	7	4.4
Cystic kidney	111	4.2	19	5.0	10	3.0	6	3.7
Hydronephrosis	339	12.9	34	8.9	37	11.1	13	8.1
Exstrophy bladder	5	0.2	0	0.0	4	1.2	0	0.0
Posterior urethral valves	17	0.6	2	0.5	2	0.6	0	0.0
LIMB & MUSCULOSKELETAL	**3525**	**133.8**	**463**	**121.0**	**381**	**114.1**	**202**	**125.6**
Scoliosis	60	2.3	10	2.6	9	2.7	4	2.5
Hip dislocation	277	10.5	29	7.6	31	9.3	21	13.1
Clubfoot	817	31.0	128	33.4	115	34.5	48	29.8
Polydactyly	475	18.0	43	11.2	55	16.5	24	14.9
Syndactyly	212	8.0	36	9.4	20	6.0	13	8.1
Limb reduction deformity - upper	91	3.5	14	3.7	9	2.7	3	1.9
Limb reduction deformity - lower	41	1.6	3	0.8	3	0.9	4	2.5
Craniosynostosis	135	5.1	19	5.0	12	3.6	7	4.4
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	12	0.5	3	0.8	1	0.3	1	0.6
Spina bifida occulta	6	0.2	3	0.8	1	0.3	2	1.2
Dwarfism	73	2.8	12	3.1	6	1.8	2	1.2
Osteogenesis imperfecta	9	0.3	2	0.5	2	0.6	2	1.2

TABLE 6. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY COUNTY, HAWAII, 1986-2003

DIAGNOSIS	HONOLULU		HAWAII		MAUI		KAUAI	
	263,409		38,268		33,380		16,081	
	N	Rate	N	Rate	N	Rate	N	Rate
Diaphragmatic hernia	65	2.5	6	1.6	7	2.1	4	2.5
Abdominal wall defects	218	8.3	31	8.1	30	9.0	14	8.7
Omphalocele	69	2.6	8	2.1	11	3.3	3	1.9
Gastroschisis	85	3.2	14	3.7	10	3.0	7	4.4
Prune-belly syndrome	7	0.3	0	0.0	1	0.3	1	0.6
Poland syndrome	4	0.2	1	0.3	0	0.0	0	0.0
SKIN & INTEGUMENT	**1806**	**68.6**	**162**	**42.3**	**105**	**31.5**	**60**	**37.3**
CHROMOSOME & SYNDROME	**1443**	**54.8**	**191**	**49.9**	**149**	**44.6**	**78**	**48.5**
Down syndrome (trisomy 21)	387	14.7	55	14.4	45	13.5	20	12.4
Patau syndrome (trisomy 13)	49	1.9	10	2.6	4	1.2	3	1.9
Edwards syndrome (trisomy 18)	127	4.8	11	2.9	16	4.8	1	0.6
Cri du Chat syndrome	4	0.2	1	0.3	0	0.0	2	1.2
Wolff-Hirschorn syndrome	5	0.2	2	0.5	1	0.3	0	0.0
Autosomal anomalies in normal infant	114	4.3	7	1.8	5	1.5	8	5.0
Triploidy	34	1.3	1	0.3	1	0.3	2	1.2
Turner syndrome (45,X)	114	4.3	10	2.6	7	2.1	4	2.5
Klinefelter syndrome (47,XXY)	37	1.4	4	1.0	6	1.8	3	1.9
Absent spleen/Asplenia	7	0.3	2	0.5	1	0.3	0	0.0
Accessory spleen	27	1.0	6	1.6	4	1.2	2	1.2
Situs inversus	25	0.9	5	1.3	2	0.6	1	0.6
Conjoined twins	10	0.4	2	0.5	0	0.0	0	0.0
Tuberous sclerosis	12	0.5	0	0.0	1	0.3	0	0.0
Amniotic band syndrome	57	2.2	7	1.8	7	2.1	5	3.1
DiGeorge syndrome	14	0.5	5	1.3	2	0.6	0	0.0
Pierre-Robin sequence	20	0.8	5	1.3	4	1.2	2	1.2
Werdnig-Hoffman syndrome	10	0.4	1	0.3	3	0.9	0	0.0
Wolff-Parkinson-White syndrome	14	0.5	1	0.3	3	0.9	1	0.6
NEOPLASMS	**281**	**10.7**	**30**	**7.8**	**26**	**7.8**	**12**	**7.5**
CONGENITAL INFECTION	**200**	**7.6**	**24**	**6.3**	**17**	**5.1**	**20**	**12.4**
MATERNAL SUBSTANCE ABUSE	**2279**	**86.5**	**304**	**79.4**	**175**	**52.4**	**96**	**59.7**
Fetal alcohol syndrome	48	1.8	28	7.3	4	1.2	8	5.0
Illicit drugs	2255	85.6	285	74.5	173	51.8	91	56.6
Marijuana	697	26.5	121	31.6	62	18.6	39	24.3
Cocaine	382	14.5	109	28.5	51	15.3	31	19.3
Methamphetamine/Amphetamine	1590	60.4	104	27.2	77	23.1	43	26.7
TOTAL	**14309**	**543.2**	**1759**	**459.7**	**1447**	**433.5**	**780**	**485.0**

*birth weight >2499 grams.

FIGURE 6. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY COUNTY, HAWAII, 1986-2003

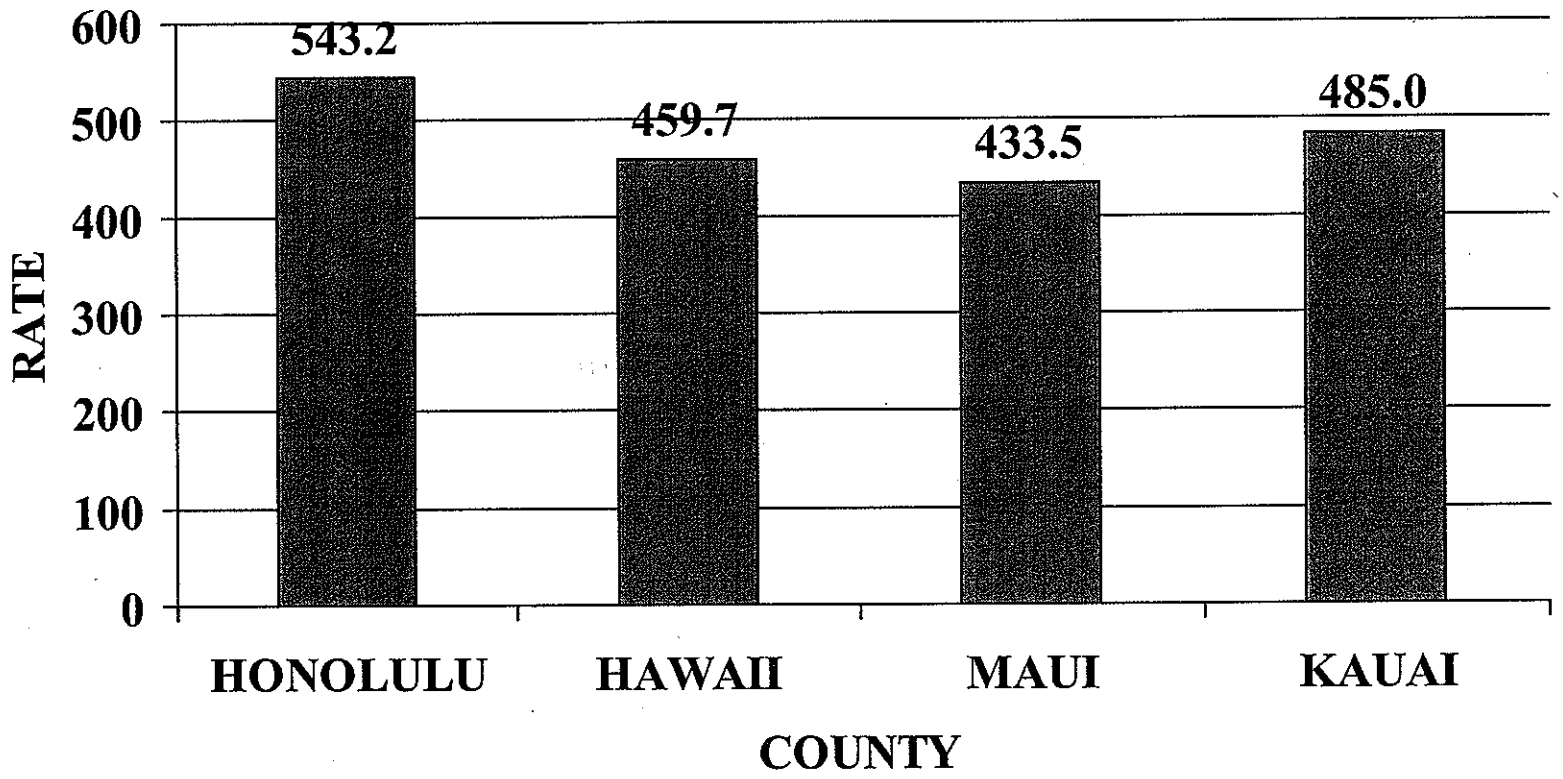


TABLE 7. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY URBANITY, HAWAII, 1986-2003

DIAGNOSIS	URBAN* 99,122		RURAL** 234,118	
	N	Rate	N	Rate
BRAIN & NERVOUS SYSTEM	**401**	**40.5**	**984**	**42.0**
Neural tube defects	96	9.7	225	9.6
Anencephaly	32	3.2	85	3.6
Spina bifida	47	4.7	98	4.2
Encephalocele	19	1.9	49	2.1
Microcephaly	93	9.4	246	10.5
Holoprosencephaly	11	1.1	30	1.3
Hydrocephaly (w/o spina bifida)	95	9.6	274	11.7
EYE, EAR, FACE, & NECK	**525**	**53.0**	**1237**	**52.8**
EYE	188	19.0	480	20.5
Anophthalmia	5	0.5	8	0.3
Microphthalmia	34	3.4	66	2.8
Cataract	8	0.8	34	1.5
EAR	371	37.4	870	37.2
Absent auditory canal/ear	12	1.2	25	1.1
Microtia	40	4.0	80	3.4
CARDIAC & CIRCULATORY	**1939**	**195.6**	**4409**	**188.3**
Common truncus	10	1.0	13	0.6
Transposition of the great vessels	34	3.4	110	4.7
Tetralogy of Fallot	38	3.8	91	3.9
Single ventricle	6	0.6	22	0.9
Ventricular septal defect	521	52.6	1085	46.3
Atrial septal defect	680	68.6	1645	70.3
Endocardial cushion defect	22	2.2	55	2.3
Pulmonary valve atresia/stenosis	98	9.9	226	9.7
Tricuspid valve atresia/stenosis	16	1.6	38	1.6
Ebstein's anomaly	6	0.6	12	0.5
Aortic valve atresia/stenosis	13	1.3	37	1.6
Mitral valve atresia/stenosis	7	0.7	30	1.3
Hypoplastic left heart syndrome	12	1.2	41	1.8
Dextrocardia	20	2.0	33	1.4
Patent ductus arteriosus***	836	84.3	1849	79.0
Coarctation of the aorta	18	1.8	79	3.4
Aorta stenosis	1	0.1	1	0.0
Pulmonary artery atresia/stenosis	200	20.2	433	18.5
Total anomalous pulmonary venous return	11	1.1	21	0.9
Partial anomalous pulmonary venous	4	0.4	12	0.5
Persistent fetal circulation	67	6.8	134	5.7
RESPIRATORY	**257**	**25.9**	**635**	**27.1**

TABLE 7. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY URBANITY, HAWAII, 1986-2003

DIAGNOSIS	URBAN* 99,122		RURAL** 234,118	
	N	Rate	N	Rate
Choanal atresia/stenosis	15	1.5	35	1.5
Lung cysts	10	1.0	18	0.8
Hypoplastic lung	63	6.4	135	5.8
OROFACIAL & GASTROINTESTINAL	**600**	**60.5**	**1446**	**61.8**
Oral clefts	206	20.8	464	19.8
Cleft palate (isolated)	67	6.8	178	7.6
Cleft lip (±cleft palate)	139	14.0	287	12.3
TE fistula/Esophageal atresia	18	1.8	53	2.3
Pyloric stenosis	73	7.4	198	8.5
Atresia/stenosis small intestine	25	2.5	66	2.8
Anal atresia/stenosis	52	5.2	116	5.0
Hirschsprung's disease	32	3.2	51	2.2
Malrotation	26	2.6	67	2.9
Biliary atresia	12	1.2	22	0.9
GENITAL & URINARY	**1125**	**113.5**	**2599**	**111.0**
Ovarian cyst	5	0.5	13	0.6
Undescended testicle***	363	36.6	770	32.9
Hypospadias/Epispadias/Chordee	296	29.9	830	35.5
Ambiguous genitalia	4	0.4	11	0.5
Absent/hypoplastic kidney	51	5.1	109	4.7
Cystic kidney	50	5.0	96	4.1
Hydronephrosis	147	14.8	275	11.7
Exstrophy bladder	0	0.0	9	0.4
Posterior urethral valves	8	0.8	13	0.6
LIMB & MUSCULOSKELETAL	**1350**	**136.2**	**3223**	**137.7**
Scoliosis	24	2.4	59	2.5
Hip dislocation	101	10.2	257	11.0
Clubfoot	311	31.4	798	34.1
Polydactyly	177	17.9	421	18.0
Syndactyly	86	8.7	195	8.3
Limb reduction deformity - upper	32	3.2	85	3.6
Limb reduction deformity - lower	16	1.6	35	1.5
Craniosynostosis	45	4.5	128	5.5
Crouzon's disease	1	0.1	1	0.0
Apert syndrome	3	0.3	8	0.3
Goldenhar syndrome	8	0.8	9	0.4
Spina bifida occulta	2	0.2	10	0.4
Dwarfism	26	2.6	67	2.9
Osteogenesis imperfecta	6	0.6	9	0.4

TABLE 7. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY URBANITY, HAWAII, 1986-2003

DIAGNOSIS	URBAN* 99,122		RURAL** 234,118	
	N	Rate	N	Rate
Diaphragmatic hernia	24	2.4	58	2.5
Abdominal wall defects	76	7.7	217	9.3
Omphalocele	27	2.7	64	2.7
Gastroschisis	29	2.9	87	3.7
Prune-belly syndrome	1	0.1	8	0.3
Poland syndrome	1	0.1	4	0.2
SKIN & INTEGUMENT	**733**	**73.9**	**1399**	**59.8**
CHROMOSOME & SYNDROME	**607**	**61.2**	**1256**	**53.6**
Down syndrome (trisomy 21)	175	17.7	332	14.2
Patau syndrome (trisomy 13)	23	2.3	43	1.8
Edwards syndrome (trisomy 18)	66	6.7	89	3.8
Cri du Chat syndrome	3	0.3	4	0.2
Wolff-Hirschorn syndrome	3	0.3	5	0.2
Autosomal anomalies in normal infant	45	4.5	89	3.8
Triploidy	17	1.7	21	0.9
Turner syndrome (45,X)	47	4.7	88	3.8
Klinefelter syndrome (47,XXY)	16	1.6	34	1.5
Absent spleen/Asplenia	3	0.3	7	0.3
Accessory spleen	9	0.9	30	1.3
Situs inversus	12	1.2	21	0.9
Conjoined twins	4	0.4	8	0.3
Tuberous sclerosis	4	0.4	9	0.4
Amniotic band syndrome	22	2.2	54	2.3
DiGeorge syndrome	6	0.6	15	0.6
Pierre-Robin sequence	7	0.7	24	1.0
Werdnig-Hoffman syndrome	1	0.1	13	0.6
Wolff-Parkinson-White syndrome	3	0.3	16	0.7
NEOPLASMS	**113**	**11.4**	**235**	**10.0**
CONGENITAL INFECTION	**87**	**8.8**	**174**	**7.4**
MATERNAL SUBSTANCE ABUSE	**714**	**72.0**	**2136**	**91.2**
Fetal alcohol syndrome	25	2.5	63	2.7
Illicit drugs	700	70.6	2100	89.7
Marijuana	205	20.7	714	30.5
Cocaine	181	18.3	389	16.6
Methamphetamine/Amphetamine	446	45.0	1366	58.3
TOTAL	**5416**	**546.4**	**12874**	**549.9**

*Metropolitan Honolulu (any zip code starting with 968).

**Rest of Hawaii (any zip code starting with 967).

***Birth weight >2499 grams.

FIGURE 7. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY URBANITY, HAWAII, 1986-2003

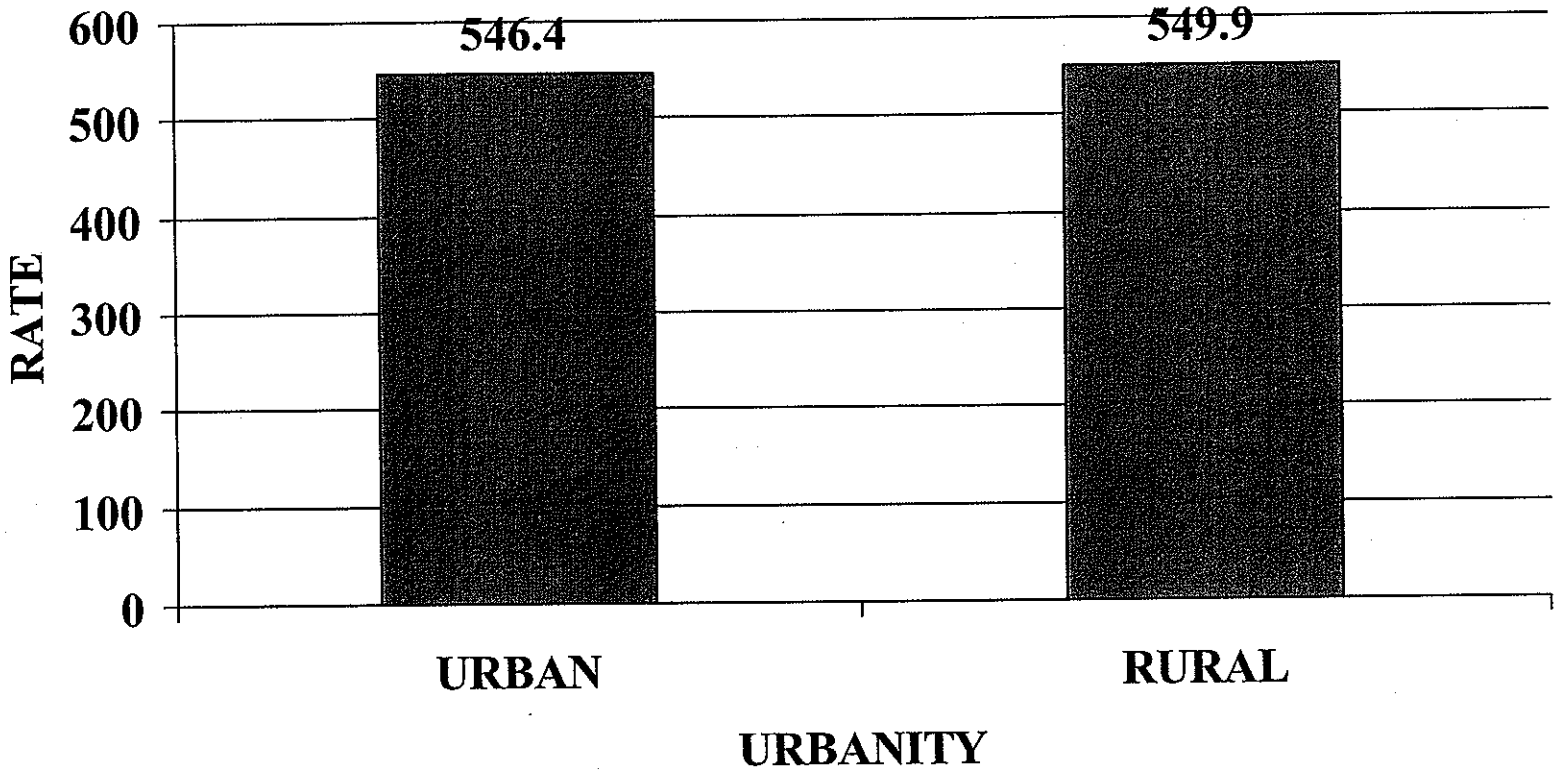


TABLE 8. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY GESTATIONAL AGE, HAWAII, 1986-2003*

DIAGNOSIS	≤19		20-27		28-30		31-33		34-37		38-40		≥41	
	157		1,721		1,682		4,130		46,017		219,668		46,667	
	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
BRAIN & NERVOUS SYSTEM	**0**	**0.0**	**55**	**319.6**	**60**	**356.7**	**89**	**215.5**	**355**	**77.1**	**481**	**21.9**	**72**	**15.4**
Neural tube defects	0	0.0	3	17.4	1	5.9	7	16.9	65	14.1	74	3.4	7	1.5
Anencephaly	0	0.0	1	5.8	1	5.9	1	2.4	10	2.2	3	0.1	3	0.6
Spina bifida	0	0.0	1	5.8	0	0.0	4	9.7	46	10.0	55	2.5	3	0.6
Encephalocele	0	0.0	1	5.8	0	0.0	2	4.8	11	2.4	17	0.8	1	0.2
Microcephaly	0	0.0	10	58.1	15	89.2	15	36.3	130	28.3	142	6.5	22	4.7
Holoprosencephaly	0	0.0	0	0.0	1	5.9	4	9.7	11	2.4	10	0.5	1	0.2
Hydrocephaly (w/o spina bifida)	0	0.0	24	139.5	23	136.7	33	79.9	92	20.0	109	5.0	17	3.6
EYE, EAR, FACE, & NECK	**0**	**0.0**	**30**	**174.3**	**38**	**225.9**	**98**	**237.3**	**484**	**105.2**	**891**	**40.6**	**134**	**28.7**
EYE	0	0.0	6	34.9	16	95.1	35	84.7	204	44.3	348	15.8	54	11.6
Anophthalmia	0	0.0	0	0.0	1	5.9	0	0.0	3	0.7	3	0.1	0	0.0
Microphthalmia	0	0.0	0	0.0	2	11.9	9	21.8	43	9.3	37	1.7	10	2.1
Cataract	0	0.0	1	5.8	1	5.9	3	7.3	9	2.0	24	1.1	6	1.3
EAR	0	0.0	26	151.1	21	124.9	77	186.4	324	70.4	639	29.1	99	21.2
Absent auditory canal/ear	0	0.0	0	0.0	0	0.0	2	4.8	13	2.8	20	0.9	2	0.4
Microtia	0	0.0	3	17.4	1	5.9	4	9.7	24	5.2	75	3.4	10	2.1
CARDIAC & CIRCULATORY	**3**	**191.1**	**371**	**2155.7**	**366**	**2176.0**	**444**	**1075.1**	**1490**	**323.8**	**3058**	**139.2**	**517**	**110.8**
Common truncus	0	0.0	1	5.8	0	0.0	0	0.0	4	0.9	8	0.4	6	1.3
Transposition of the great vessels	0	0.0	0	0.0	7	41.6	3	7.3	30	6.5	80	3.6	16	3.4
Tetralogy of Fallot	0	0.0	2	11.6	2	11.9	11	26.6	29	6.3	71	3.2	9	1.9
Single ventricle	0	0.0	1	5.8	2	11.9	1	2.4	5	1.1	12	0.5	4	0.9
Ventricular septal defect	1	63.7	36	209.2	43	255.6	79	191.3	377	81.9	882	40.2	154	33.0
Atrial septal defect	1	63.7	187	1086.6	152	903.7	179	433.4	638	138.6	990	45.1	157	33.6
Endocardial cushion defect	0	0.0	1	5.8	3	17.8	4	9.7	20	4.3	35	1.6	9	1.9
Pulmonary valve atresia/stenosis	0	0.0	10	58.1	17	101.1	25	60.5	81	17.6	157	7.1	30	6.4
Tricuspid valve atresia/stenosis	0	0.0	1	5.8	0	0.0	2	4.8	14	3.0	33	1.5	4	0.9
Ebstein's anomaly	0	0.0	0	0.0	2	11.9	0	0.0	3	0.7	11	0.5	0	0.0

TABLE 8. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY GESTATIONAL AGE, HAWAII, 1986-2003*

DIAGNOSIS	≤19		20-27		28-30		31-33		34-37		38-40		≥41	
	157		1,721		1,682		4,130		46,017		219,668		46,667	
	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Aortic valve atresia/stenosis	0	0.0	2	11.6	1	5.9	2	4.8	15	3.3	25	1.1	4	0.9
Mitral valve atresia/stenosis	0	0.0	0	0.0	0	0.0	3	7.3	12	2.6	20	0.9	2	0.4
Hypoplastic left heart syndrome	0	0.0	1	5.8	1	5.9	3	7.3	10	2.2	25	1.1	2	0.4
Dextrocardia	0	0.0	1	5.8	0	0.0	3	7.3	13	2.8	26	1.2	8	1.7
Patent ductus arteriosus**	2	127.4	1	5.8	4	23.8	24	58.1	614	133.4	1762	80.2	285	61.1
Coarctation of the aorta	0	0.0	0	0.0	3	17.8	5	12.1	24	5.2	52	2.4	10	2.1
Aorta stenosis	0	0.0	0	0.0	1	5.9	0	0.0	0	0.0	1	0.0	0	0.0
Pulmonary artery atresia/stenosis	0	0.0	53	308.0	81	481.6	105	254.2	182	39.6	175	8.0	33	7.1
Total anomalous pulmonary venous return	0	0.0	0	0.0	0	0.0	1	2.4	6	1.3	22	1.0	3	0.6
Partial anomalous pulmonary venous	0	0.0	0	0.0	0	0.0	0	0.0	5	1.1	10	0.5	1	0.2
Persistent fetal circulation	0	0.0	3	17.4	5	29.7	7	16.9	41	8.9	100	4.6	44	9.4
RESPIRATORY	**0**	**0.0**	**37**	**215.0**	**31**	**184.3**	**78**	**188.9**	**244**	**53.0**	**383**	**17.4**	**70**	**15.0**
Choanal atresia/stenosis	0	0.0	1	5.8	0	0.0	3	7.3	8	1.7	32	1.5	6	1.3
Lung cysts	0	0.0	0	0.0	0	0.0	3	7.3	5	1.1	15	0.7	2	0.4
Hypoplastic lung	0	0.0	22	127.8	17	101.1	42	101.7	59	12.8	35	1.6	7	1.5
OROFACIAL & GASTROINTESTINAL	**0**	**0.0**	**32**	**185.9**	**37**	**220.0**	**105**	**254.2**	**525**	**114.1**	**1072**	**48.8**	**182**	**39.0**
Oral clefts	0	0.0	4	23.2	8	47.6	26	63.0	139	30.2	377	17.2	63	13.5
Cleft palate (isolated)	0	0.0	3	17.4	1	5.9	9	21.8	57	12.4	141	6.4	20	4.3
Cleft lip (±cleft palate)	0	0.0	1	5.8	7	41.6	17	41.2	82	17.8	236	10.7	43	9.2
TE fistula/Esophageal atresia	0	0.0	1	5.8	0	0.0	11	26.6	20	4.3	24	1.1	11	2.4
Pyloric stenosis	0	0.0	1	5.8	0	0.0	6	14.5	55	12.0	175	8.0	26	5.6
Atresia/stenosis small intestine	0	0.0	1	5.8	3	17.8	12	29.1	43	9.3	28	1.3	2	0.4
Anal atresia/stenosis	0	0.0	1	5.8	9	53.5	15	36.3	50	10.9	77	3.5	7	1.5
Hirschsprung's disease	0	0.0	3	17.4	0	0.0	3	7.3	19	4.1	47	2.1	11	2.4
Malrotation	0	0.0	1	5.8	1	5.9	8	19.4	32	7.0	44	2.0	3	0.6
Biliary atresia	0	0.0	0	0.0	2	11.9	1	2.4	7	1.5	22	1.0	3	0.6
GENITAL & URINARY	**1**	**63.7**	**98**	**569.4**	**97**	**576.7**	**175**	**423.7**	**848**	**184.3**	**2102**	**95.7**	**320**	**68.6**

TABLE 8. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY GESTATIONAL AGE, HAWAII, 1986-2003*

DIAGNOSIS	≤19		20-27		28-30		31-33		34-37		38-40		≥41	
	157		1,721		1,682		4,130		46,017		219,668		46,667	
	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Ovarian cyst	0	0.0	1	5.8	1	5.9	1	2.4	1	0.2	9	0.4	5	1.1
Undescended testicle**	0	0.0	0	0.0	0	0.0	8	19.4	218	47.4	771	35.1	138	29.6
Hypospadias/Epispadias/Chordee	1	63.7	12	69.7	23	136.7	60	145.3	238	51.7	693	31.5	93	19.9
Ambiguous genitalia	0	0.0	2	11.6	0	0.0	0	0.0	1	0.2	5	0.2	1	0.2
Absent/hypoplastic kidney	0	0.0	4	23.2	8	47.6	18	43.6	49	10.6	46	2.1	9	1.9
Cystic kidney	0	0.0	4	23.2	8	47.6	17	41.2	43	9.3	57	2.6	6	1.3
Hydronephrosis	0	0.0	15	87.2	15	89.2	30	72.6	130	28.3	202	9.2	19	4.1
Exstrophy bladder	0	0.0	0	0.0	0	0.0	0	0.0	3	0.7	4	0.2	1	0.2
Posterior urethral valves	0	0.0	0	0.0	0	0.0	2	4.8	5	1.1	8	0.4	1	0.2
LIMB & MUSCULOSKELETAL	**1**	**63.7**	**47**	**273.1**	**72**	**428.1**	**154**	**372.9**	**1015**	**220.6**	**2590**	**117.9**	**498**	**106.7**
Scoliosis	0	0.0	4	23.2	2	11.9	5	12.1	25	5.4	27	1.2	6	1.3
Hip dislocation	0	0.0	2	11.6	3	17.8	8	19.4	45	9.8	224	10.2	76	16.3
Clubfoot	1	63.7	10	58.1	13	77.3	27	65.4	235	51.1	681	31.0	115	24.6
Polydactyly	0	0.0	1	5.8	5	29.7	20	48.4	98	21.3	391	17.8	65	13.9
Syndactyly	0	0.0	5	29.1	5	29.7	18	43.6	66	14.3	137	6.2	39	8.4
Limb reduction deformity - upper	0	0.0	8	46.5	2	11.9	7	16.9	29	6.3	42	1.9	14	3.0
Limb reduction deformity - lower	0	0.0	5	29.1	0	0.0	3	7.3	13	2.8	18	0.8	5	1.1
Craniosynostosis	0	0.0	1	5.8	1	5.9	3	7.3	44	9.6	96	4.4	25	5.4
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	1.1	6	0.3	0	0.0
Goldenhar syndrome	0	0.0	1	5.8	0	0.0	1	2.4	5	1.1	11	0.5	1	0.2
Spina bifida occulta	0	0.0	0	0.0	1	5.9	0	0.0	1	0.2	7	0.3	1	0.2
Dwarfism	0	0.0	3	17.4	4	23.8	4	9.7	25	5.4	38	1.7	9	1.9
Osteogenesis imperfecta	0	0.0	0	0.0	1	5.9	0	0.0	5	1.1	6	0.3	2	0.4
Diaphragmatic hernia	0	0.0	3	17.4	2	11.9	5	12.1	17	3.7	39	1.8	7	1.5
Abdominal wall defects	0	0.0	5	29.1	6	35.7	17	41.2	118	25.6	74	3.4	14	3.0
Omphalocele	0	0.0	2	11.6	3	17.8	4	9.7	26	5.7	17	0.8	4	0.9

TABLE 8. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY GESTATIONAL AGE, HAWAII, 1986-2003*

DIAGNOSIS	≤19		20-27		28-30		31-33		34-37		38-40		≥41	
	157		1,721		1,682		4,130		46,017		219,668		46,667	
	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Gastroschisis	0	0.0	1	5.8	1	5.9	10	24.2	74	16.1	14	0.6	2	0.4
Prune-belly syndrome	0	0.0	1	5.8	1	5.9	1	2.4	4	0.9	1	0.0	0	0.0
Poland syndrome	0	0.0	0	0.0	0	0.0	1	2.4	1	0.2	3	0.1	0	0.0
SKIN & INTEGUMENT	**0**	**0.0**	**10**	**58.1**	**21**	**124.9**	**55**	**133.2**	**560**	**121.7**	**1278**	**58.2**	**198**	**42.4**
CHROMOSOME & SYNDROME	**4**	**254.8**	**17**	**98.8**	**21**	**124.9**	**61**	**147.7**	**344**	**74.8**	**600**	**27.3**	**82**	**17.6**
Down syndrome (trisomy 21)	2	127.4	1	5.8	4	23.8	9	21.8	118	25.6	157	7.1	9	1.9
Patau syndrome (trisomy 13)	0	0.0	0	0.0	2	11.9	1	2.4	14	3.0	6	0.3	2	0.4
Edwards syndrome (trisomy 18)	0	0.0	2	11.6	3	17.8	9	21.8	16	3.5	17	0.8	5	1.1
Cri du Chat syndrome	0	0.0	0	0.0	0	0.0	0	0.0	1	0.2	1	0.0	2	0.4
Wolff-Hirschorn syndrome	0	0.0	0	0.0	0	0.0	1	2.4	4	0.9	2	0.1	0	0.0
Autosomal anomalies in normal infant	0	0.0	1	5.8	1	5.9	2	4.8	23	5.0	87	4.0	10	2.1
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)	2	127.4	1	5.8	0	0.0	0	0.0	5	1.1	23	1.0	1	0.2
Klinefelter syndrome (47,XXY)	0	0.0	0	0.0	0	0.0	0	0.0	7	1.5	24	1.1	1	0.2
Absent spleen/Asplenia	0	0.0	0	0.0	0	0.0	1	2.4	3	0.7	3	0.1	0	0.0
Accessory spleen	0	0.0	1	5.8	3	17.8	2	4.8	12	2.6	9	0.4	2	0.4
Situs inversus	0	0.0	0	0.0	0	0.0	2	4.8	9	2.0	20	0.9	4	0.9
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	0	0.0	3	0.7	9	0.4	1	0.2
Amniotic band syndrome	0	0.0	5	29.1	3	17.8	4	9.7	18	3.9	21	1.0	7	1.5
DiGeorge syndrome	0	0.0	0	0.0	0	0.0	0	0.0	5	1.1	13	0.6	3	0.6
Pierre-Robin sequence	0	0.0	0	0.0	0	0.0	2	4.8	5	1.1	21	1.0	2	0.4
Werdnig-Hoffman syndrome	0	0.0	0	0.0	2	11.9	2	4.8	2	0.4	4	0.2	1	0.2
Wolff-Parkinson-White syndrome	0	0.0	0	0.0	1	5.9	0	0.0	1	0.2	14	0.6	2	0.4
NEOPLASMS	**1**	**63.7**	**4**	**23.2**	**2**	**11.9**	**6**	**14.5**	**48**	**10.4**	**132**	**6.0**	**20**	**4.3**
CONGENITAL INFECTION	**0**	**0.0**	**19**	**110.4**	**22**	**130.8**	**16**	**38.7**	**66**	**14.3**	**107**	**4.9**	**18**	**3.9**
MATERNAL SUBSTANCE ABUSE	**2**	**127.4**	**53**	**308.0**	**55**	**327.0**	**125**	**302.7**	**825**	**179.3**	**1518**	**69.1**	**178**	**38.1**

TABLE 8. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY GESTATIONAL AGE, HAWAII, 1986-2003*

DIAGNOSIS	≤19		20-27		28-30		31-33		34-37		38-40		≥41	
	157		1,721		1,682		4,130		46,017		219,668		46,667	
	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Fetal alcohol syndrome	0	0.0	2	11.6	4	23.8	3	7.3	22	4.8	45	2.0	8	1.7
Illicit drugs	2	127.4	52	302.1	53	315.1	124	300.2	813	176.7	1487	67.7	176	37.7
Marijuana	0	0.0	22	127.8	18	107.0	53	128.3	219	47.6	485	22.1	87	18.6
Cocaine	0	0.0	11	63.9	11	65.4	32	77.5	167	36.3	300	13.7	26	5.6
Methamphetamine/Amphetamine	2	127.4	26	151.1	30	178.4	80	193.7	587	127.6	944	43.0	94	20.1
TOTAL	**8**	**509.6**	**443**	**2574.1**	**458**	**2722.9**	**746**	**1806.3**	**3921**	**852.1**	**9895**	**450.5**	**1605**	**343.9**

*Live births.

**Birth weight >2499 grams.

FIGURE 8. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY GESTATIONAL AGE, HAWAII, 1986-2003

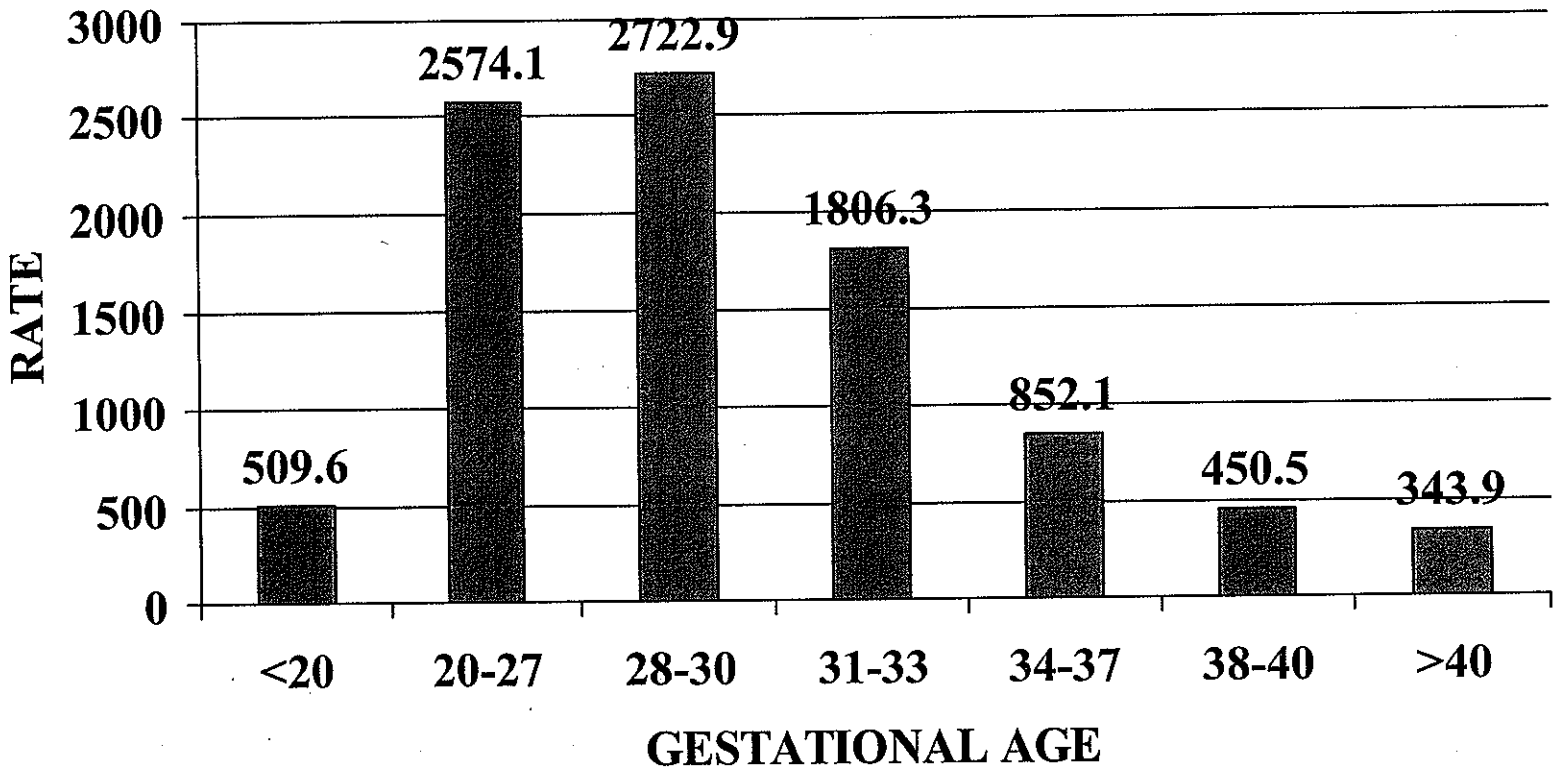
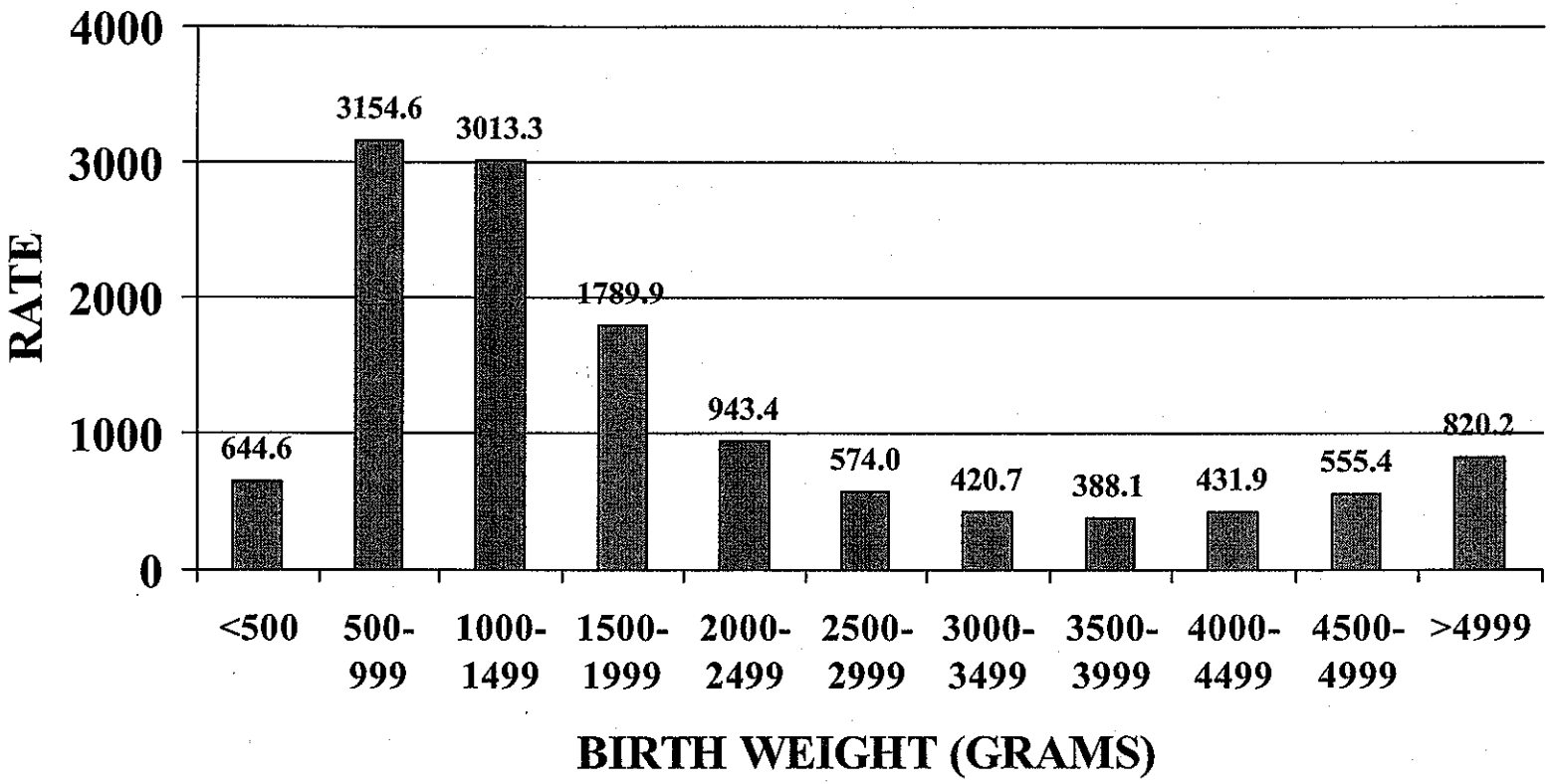
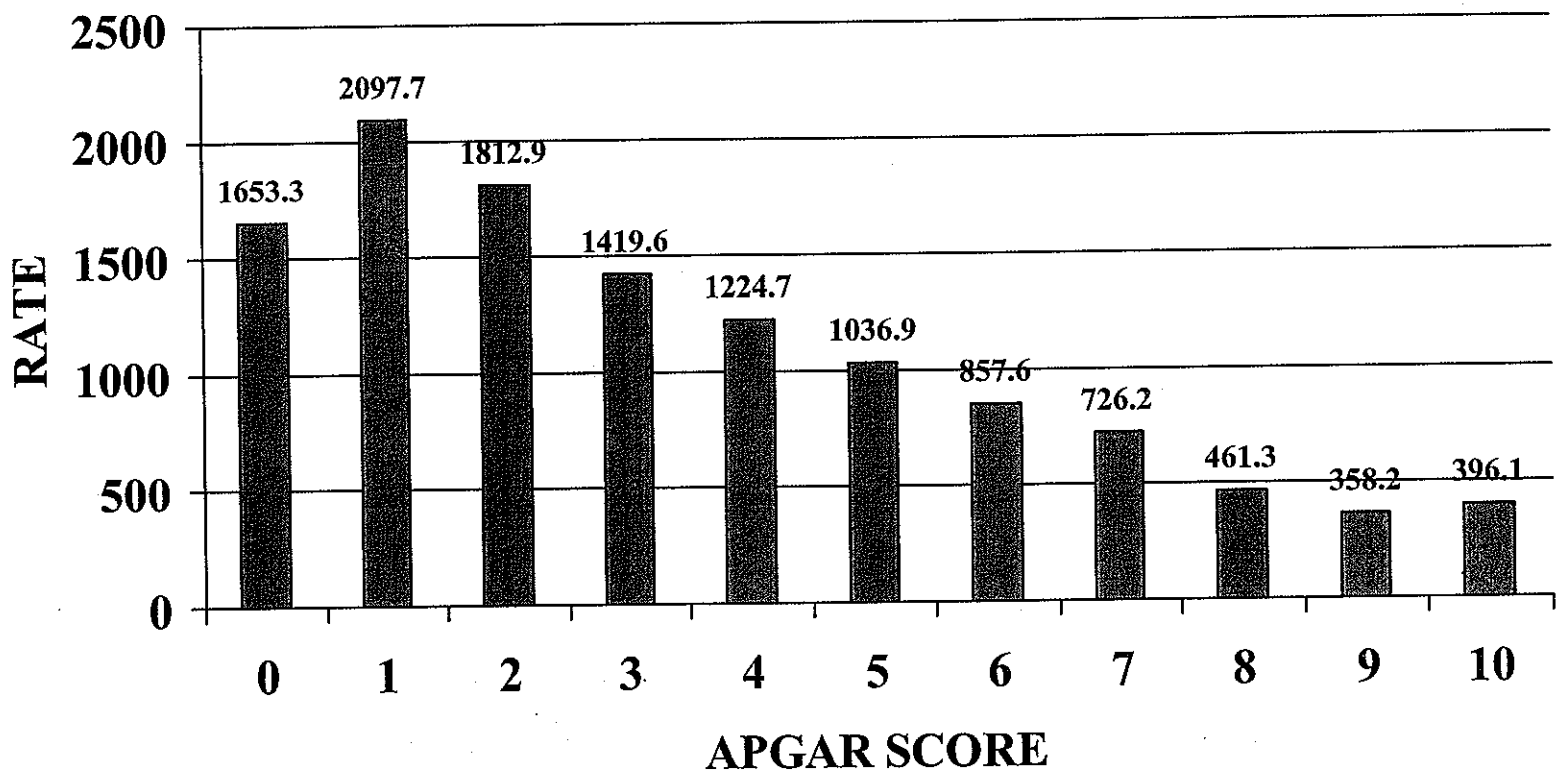


FIGURE 9. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY BIRTH WEIGHT, HAWAII, 1986-2003



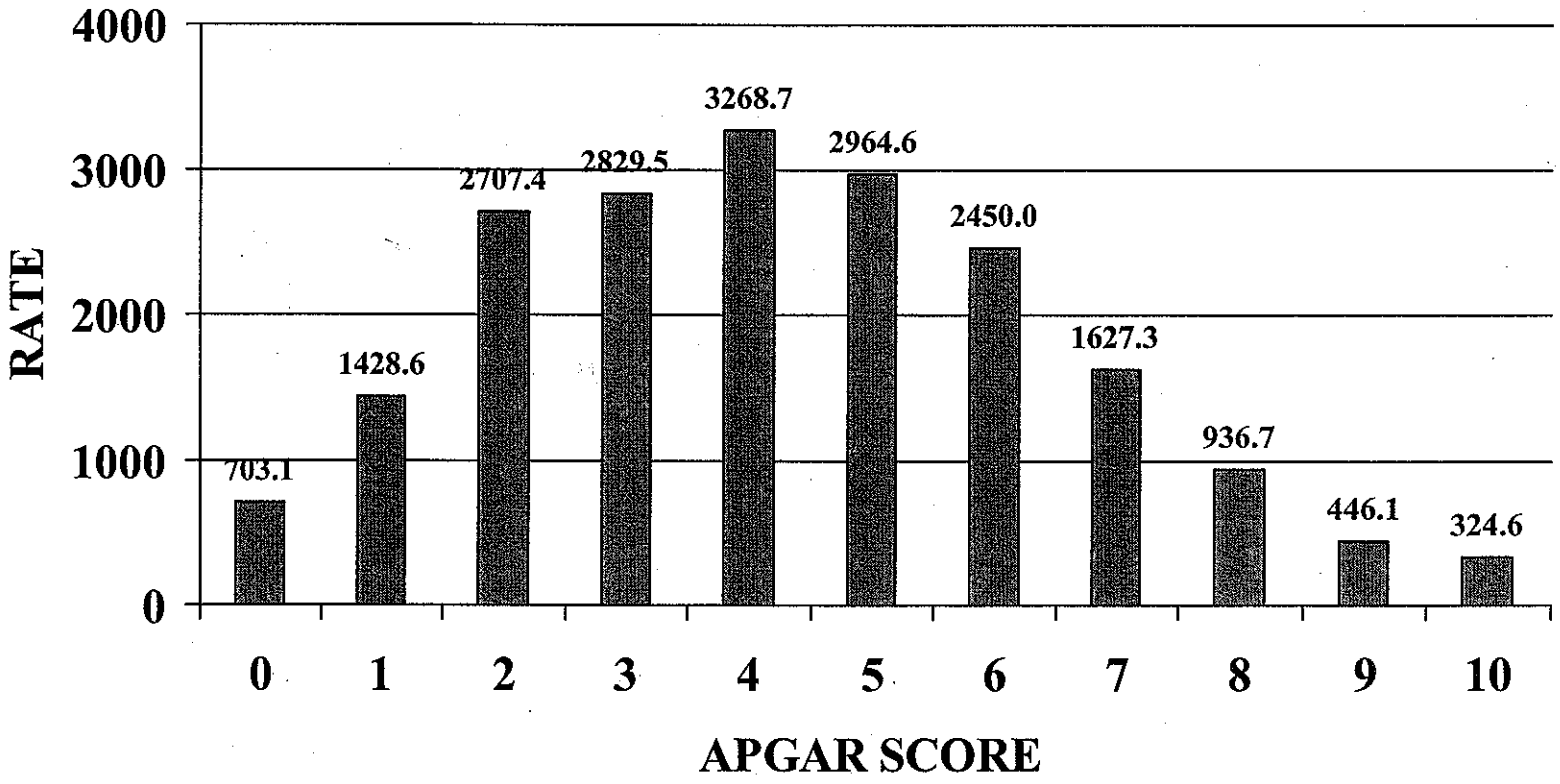
*Live births only

FIGURE 10. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY 1 MINUTE APGAR SCORES, HAWAII, 1986-2003



*Live births only

FIGURE 11. RATE OF BIRTH DEFECTS PER 10,000 LIVE BIRTHS BY 5 MINUTE APGAR SCORES, HAWAII, 1986-2003



*Live births only

TABLE 9. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY MATERNAL AGE, HAWAII, 1986-2003

DIAGNOSIS	≤19		20-24		25-29		30-34		35-39		≥40	
	33,261		86,574		92,449		76,409		37,668		7,828	
	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
BRAIN & NERVOUS SYSTEM	**178**	**53.5**	**362**	**41.8**	**367**	**39.7**	**284**	**37.2**	**178**	**47.3**	**53**	**67.7**
Neural tube defects	38	11.4	71	8.2	101	10.9	70	9.2	43	11.4	10	12.8
Anencephaly	18	5.4	22	2.5	40	4.3	28	3.7	10	2.7	2	2.6
Spina bifida	17	5.1	33	3.8	39	4.2	33	4.3	28	7.4	4	5.1
Encephalocele	3	0.9	18	2.1	26	2.8	11	1.4	6	1.6	4	5.1
Microcephaly	42	12.6	103	11.9	71	7.7	72	9.4	47	12.5	10	12.8
Holoprosencephaly	7	2.1	11	1.3	7	0.8	10	1.3	4	1.1	3	3.8
Hydrocephaly (w/o spina bifida)	45	13.5	101	11.7	123	13.3	66	8.6	39	10.4	10	12.8
EYE, EAR, FACE, & NECK	**169**	**50.8**	**437**	**50.5**	**434**	**46.9**	**397**	**52.0**	**255**	**67.7**	**106**	**135.4**
EYE	64	19.2	180	20.8	165	17.8	131	17.1	98	26.0	48	61.3
Anophthalmia	1	0.3	4	0.5	5	0.5	3	0.4	0	0.0	0	0.0
Microphthalmia	11	3.3	22	2.5	31	3.4	23	3.0	15	4.0	2	2.6
Cataract	6	1.8	13	1.5	14	1.5	6	0.8	3	0.8	2	2.6
EAR	124	37.3	305	35.2	310	33.5	285	37.3	176	46.7	64	81.8
Absent auditory canal/ear	5	1.5	11	1.3	8	0.9	8	1.0	4	1.1	1	1.3
Microtia	10	3.0	33	3.8	26	2.8	30	3.9	19	5.0	3	3.8
CARDIAC & CIRCULATORY	**699**	**210.2**	**1591**	**183.8**	**1646**	**178.0**	**1456**	**190.6**	**811**	**215.3**	**233**	**297.6**
Common truncus	1	0.3	7	0.8	6	0.6	4	0.5	5	1.3	1	1.3
Transposition of the great vessels	13	3.9	39	4.5	39	4.2	35	4.6	14	3.7	7	8.9
Tetralogy of Fallot	15	4.5	36	4.2	34	3.7	26	3.4	14	3.7	6	7.7
Single ventricle	5	1.5	7	0.8	6	0.6	5	0.7	4	1.1	1	1.3
Ventricular septal defect	145	43.6	395	45.6	417	45.1	382	50.0	217	57.6	67	85.6
Atrial septal defect	265	79.7	545	63.0	592	64.0	520	68.1	324	86.0	110	140.5
Endocardial cushion defect	9	2.7	15	1.7	19	2.1	13	1.7	20	5.3	5	6.4
Pulmonary valve atresia/stenosis	41	12.3	88	10.2	80	8.7	69	9.0	39	10.4	13	16.6
Tricuspid valve atresia/stenosis	9	2.7	16	1.8	10	1.1	12	1.6	6	1.6	3	3.8
Ebstein's anomaly	0	0.0	3	0.3	7	0.8	5	0.7	2	0.5	1	1.3

TABLE 9. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY MATERNAL AGE, HAWAII, 1986-2003

DIAGNOSIS	≤19		20-24		25-29		30-34		35-39		≥40	
	33,261		86,574		92,449		76,409		37,668		7,828	
	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Aortic valve atresia/stenosis	5	1.5	9	1.0	15	1.6	11	1.4	10	2.7	1	1.3
Mitral valve atresia/stenosis	3	0.9	11	1.3	9	1.0	3	0.4	10	2.7	1	1.3
Hypoplastic left heart syndrome	5	1.5	16	1.8	12	1.3	7	0.9	9	2.4	4	5.1
Dextrocardia	3	0.9	15	1.7	18	1.9	7	0.9	8	2.1	2	2.6
Patent ductus arteriosus*	273	82.1	663	76.6	727	78.6	614	80.4	322	85.5	100	127.7
Coarctation of the aorta	9	2.7	23	2.7	35	3.8	17	2.2	11	2.9	2	2.6
Aorta stenosis	0	0.0	0	0.0	0	0.0	1	0.1	1	0.3	0	0.0
Pulmonary artery atresia/stenosis	84	25.3	147	17.0	160	17.3	124	16.2	92	24.4	29	37.0
Total anomalous pulmonary venous return	2	0.6	4	0.5	10	1.1	11	1.4	5	1.3	0	0.0
Partial anomalous pulmonary venous	1	0.3	5	0.6	4	0.4	7	0.9	0	0.0	0	0.0
Persistent fetal circulation	9	2.7	51	5.9	53	5.7	52	6.8	29	7.7	5	6.4
RESPIRATORY	**102**	**30.7**	**255**	**29.5**	**231**	**25.0**	**173**	**22.6**	**103**	**27.3**	**44**	**56.2**
Choanal atresia/stenosis	3	0.9	13	1.5	11	1.2	13	1.7	8	2.1	2	2.6
Lung cysts	6	1.8	8	0.9	6	0.6	6	0.8	1	0.3	0	0.0
Hypoplastic lung	28	8.4	60	6.9	60	6.5	34	4.4	23	6.1	4	5.1
OROFACIAL & GASTROINTESTINAL	**215**	**64.6**	**532**	**61.5**	**563**	**60.9**	**450**	**58.9**	**240**	**63.7**	**80**	**102.2**
Oral clefts	63	18.9	169	19.5	189	20.4	150	19.6	81	21.5	28	35.8
Cleft palate (isolated)	24	7.2	59	6.8	62	6.7	62	8.1	33	8.8	9	11.5
Cleft lip (±cleft palate)	39	11.7	111	12.8	127	13.7	88	11.5	48	12.7	19	24.3
TE fistula/Esophageal atresia	7	2.1	14	1.6	24	2.6	19	2.5	3	0.8	4	5.1
Pyloric stenosis	41	12.3	66	7.6	65	7.0	66	8.6	27	7.2	7	8.9
Atresia/stenosis small intestine	9	2.7	25	2.9	24	2.6	19	2.5	14	3.7	5	6.4
Anal atresia/stenosis	20	6.0	38	4.4	51	5.5	36	4.7	22	5.8	4	5.1
Hirschsprung's disease	13	3.9	18	2.1	26	2.8	16	2.1	10	2.7	0	0.0
Malrotation	12	3.6	32	3.7	28	3.0	19	2.5	3	0.8	3	3.8
Biliary atresia	4	1.2	14	1.6	9	1.0	7	0.9	1	0.3	1	1.3
GENITAL & URINARY	**394**	**118.5**	**1056**	**122.0**	**1005**	**108.7**	**787**	**103.0**	**436**	**115.7**	**102**	**130.3**

TABLE 9. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY MATERNAL AGE, HAWAII, 1986-2003

DIAGNOSIS	≤19		20-24		25-29		30-34		35-39		≥40	
	33,261		86,574		92,449		76,409		37,668		7,828	
	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Ovarian cyst	1	0.3	6	0.7	4	0.4	4	0.5	3	0.8	0	0.0
Undescended testicle*	140	42.1	344	39.7	291	31.5	236	30.9	109	28.9	17	21.7
Hypospadias/Epispadias/Chordee	114	34.3	302	34.9	316	34.2	221	28.9	153	40.6	29	37.0
Ambiguous genitalia	1	0.3	5	0.6	2	0.2	3	0.4	4	1.1	0	0.0
Absent/hypoplastic kidney	17	5.1	45	5.2	55	5.9	31	4.1	15	4.0	4	5.1
Cystic kidney	14	4.2	44	5.1	39	4.2	31	4.1	17	4.5	9	11.5
Hydronephrosis	41	12.3	120	13.9	116	12.5	95	12.4	46	12.2	15	19.2
Exstrophy bladder	0	0.0	1	0.1	2	0.2	4	0.5	1	0.3	1	1.3
Posterior urethral valves	2	0.6	8	0.9	4	0.4	6	0.8	1	0.3	1	1.3
LIMB & MUSCULOSKELETAL	**493**	**148.2**	**1242**	**143.5**	**1239**	**134.0**	**982**	**128.5**	**522**	**138.6**	**157**	**200.6**
Scoliosis	16	4.8	22	2.5	23	2.5	19	2.5	7	1.9	3	3.8
Hip dislocation	35	10.5	74	8.5	99	10.7	85	11.1	58	15.4	9	11.5
Clubfoot	143	43.0	355	41.0	287	31.0	224	29.3	96	25.5	22	28.1
Polydactyly	59	17.7	181	20.9	175	18.9	127	16.6	41	10.9	19	24.3
Syndactyly	28	8.4	83	9.6	73	7.9	60	7.9	27	7.2	14	17.9
Limb reduction deformity - upper	18	5.4	30	3.5	27	2.9	29	3.8	12	3.2	3	3.8
Limb reduction deformity - lower	5	1.5	19	2.2	10	1.1	11	1.4	4	1.1	2	2.6
Craniosynostosis	10	3.0	39	4.5	59	6.4	36	4.7	27	7.2	4	5.1
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	1.6	1	1.3
Goldenhar syndrome	3	0.9	2	0.2	5	0.5	3	0.4	5	1.3	1	1.3
Spina bifida occulta	1	0.3	1	0.1	4	0.4	2	0.3	2	0.5	2	2.6
Dwarfism	10	3.0	21	2.4	22	2.4	22	2.9	17	4.5	4	5.1
Osteogenesis imperfecta	3	0.9	6	0.7	2	0.2	4	0.5	0	0.0	0	0.0
Diaphragmatic hernia	5	1.5	21	2.4	26	2.8	15	2.0	14	3.7	4	5.1
Abdominal wall defects	48	14.4	98	11.3	67	7.2	51	6.7	25	6.6	12	15.3
Omphalocele	6	1.8	27	3.1	16	1.7	24	3.1	12	3.2	10	12.8

TABLE 9. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY MATERNAL AGE, HAWAII, 1986-2003

DIAGNOSIS	≤19		20-24		25-29		30-34		35-39		≥40	
	33,261		86,574		92,449		76,409		37,668		7,828	
	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Gastroschisis	37	11.1	47	5.4	22	2.4	8	1.0	3	0.8	1	1.3
Prune-belly syndrome	1	0.3	3	0.3	2	0.2	2	0.3	1	0.3	0	0.0
Poland syndrome	1	0.3	1	0.1	2	0.2	1	0.1	0	0.0	0	0.0
SKIN & INTEGUMENT	**226**	**67.9**	**577**	**66.6**	**563**	**60.9**	**452**	**59.2**	**256**	**68.0**	**80**	**102.2**
CHROMOSOME & SYNDROME	**107**	**32.2**	**300**	**34.7**	**328**	**35.5**	**405**	**53.0**	**524**	**139.1**	**259**	**330.9**
Down syndrome (trisomy 21)	18	5.4	60	6.9	63	6.8	104	13.6	164	43.5	111	141.8
Patau syndrome (trisomy 13)	1	0.3	8	0.9	7	0.8	18	2.4	22	5.8	11	14.1
Edwards syndrome (trisomy 18)	5	1.5	18	2.1	28	3.0	34	4.4	35	9.3	41	52.4
Cri du Chat syndrome	0	0.0	0	0.0	2	0.2	3	0.4	3	0.8	0	0.0
Wolff-Hirschorn syndrome	1	0.3	3	0.3	0	0.0	3	0.4	0	0.0	1	1.3
Autosomal anomalies in normal infant	3	0.9	5	0.6	10	1.1	40	5.2	66	17.5	13	16.6
Triploidy	2	0.6	9	1.0	5	0.5	11	1.4	9	2.4	6	7.7
Turner syndrome (45,X)	9	2.7	29	3.3	32	3.5	26	3.4	35	9.3	9	11.5
Klinefelter syndrome (47,XXY)	0	0.0	7	0.8	6	0.6	6	0.8	23	6.1	9	11.5
Absent spleen/Asplenia	1	0.3	4	0.5	2	0.2	2	0.3	2	0.5	0	0.0
Accessory spleen	9	2.7	11	1.3	9	1.0	7	0.9	4	1.1	1	1.3
Situs inversus	3	0.9	8	0.9	15	1.6	6	0.8	5	1.3	0	0.0
Conjoined twins	0	0.0	6	0.7	2	0.2	2	0.3	2	0.5	0	0.0
Tuberous sclerosis	0	0.0	5	0.6	3	0.3	4	0.5	1	0.3	0	0.0
Amniotic band syndrome	12	3.6	22	2.5	18	1.9	20	2.6	4	1.1	1	1.3
DiGeorge syndrome	1	0.3	9	1.0	4	0.4	5	0.7	2	0.5	0	0.0
Pierre-Robin sequence	7	2.1	7	0.8	12	1.3	4	0.5	1	0.3	0	0.0
Werdnig-Hoffman syndrome	1	0.3	4	0.5	5	0.5	2	0.3	1	0.3	1	1.3
Wolff-Parkinson-White syndrome	6	1.8	5	0.6	3	0.3	4	0.5	1	0.3	0	0.0
NEOPLASMS	**28**	**8.4**	**89**	**10.3**	**90**	**9.7**	**100**	**13.1**	**40**	**10.6**	**9**	**11.5**
CONGENITAL INFECTION	**42**	**12.6**	**92**	**10.6**	**51**	**5.5**	**43**	**5.6**	**30**	**8.0**	**8**	**10.2**
MATERNAL SUBSTANCE ABUSE	**389**	**117.0**	**814**	**94.0**	**787**	**85.1**	**551**	**72.1**	**269**	**71.4**	**48**	**61.3**

TABLE 9. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY MATERNAL AGE, HAWAII, 1986-2003

DIAGNOSIS	≤19		20-24		25-29		30-34		35-39		≥40	
	33,261		86,574		92,449		76,409		37,668		7,828	
	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Fetal alcohol syndrome	11	3.3	24	2.8	17	1.8	17	2.2	17	4.5	3	3.8
Illicit drugs	380	114.2	801	92.5	780	84.4	544	71.2	256	68.0	46	58.8
Marijuana	189	56.8	276	31.9	211	22.8	150	19.6	78	20.7	15	19.2
Cocaine	53	15.9	138	15.9	170	18.4	130	17.0	75	19.9	7	8.9
Methamphetamine/Amphetamine	197	59.2	545	63.0	534	57.8	360	47.1	155	41.1	26	33.2
TOTAL	**1927**	**579.4**	**4748**	**548.4**	**4826**	**522.0**	**4001**	**523.6**	**2363**	**627.3**	**639**	**816.3**

*birth weight >2499 grams.

FIGURE 12. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY MATERNAL AGE, HAWAII, 1986-2003

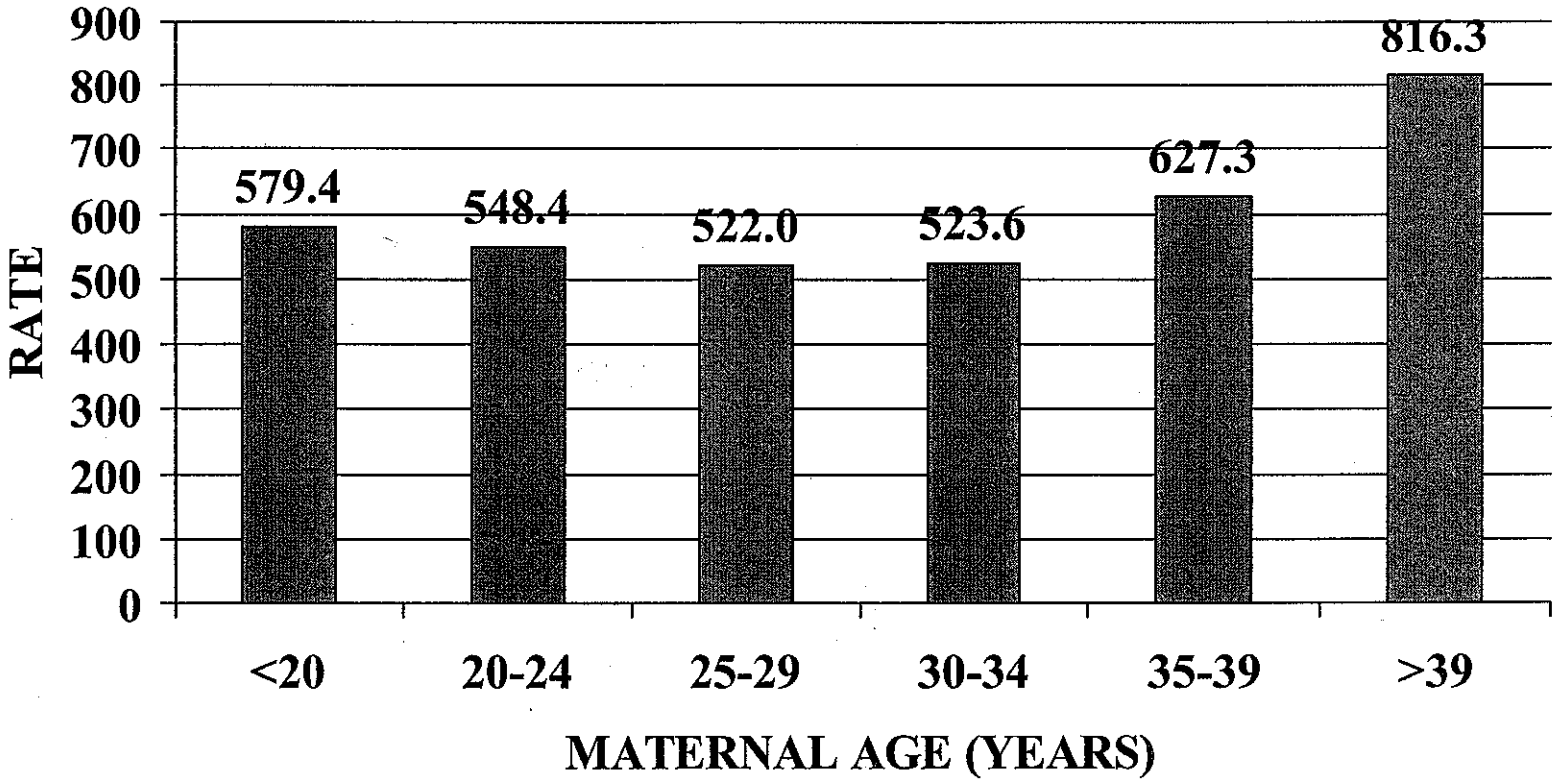


FIGURE 13. DISTRIBUTION OF BIRTH DEFECTS BY MATERNAL AGE BY 2-YEAR INTERVALS, HAWAII, 1986-2003

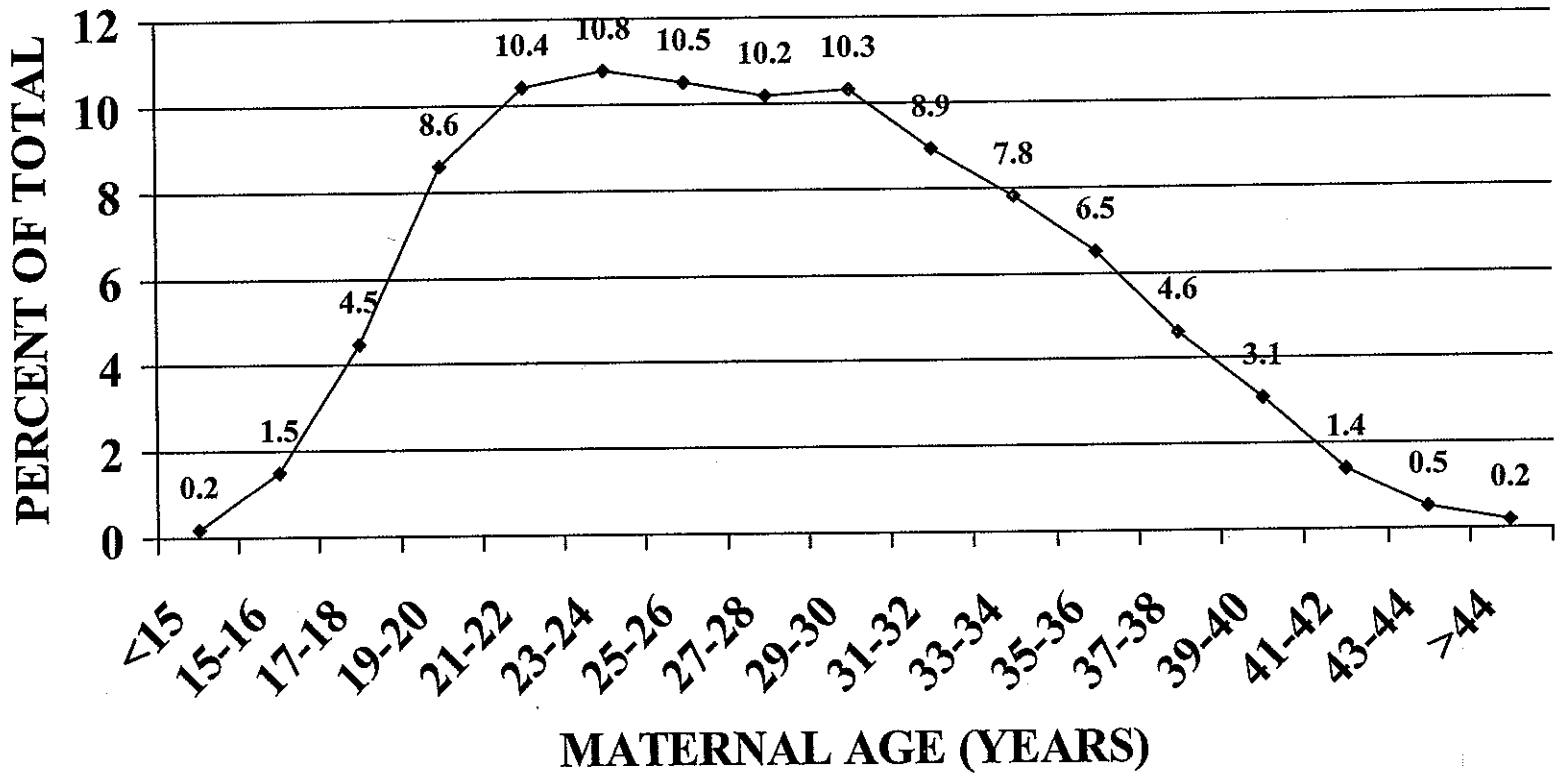


TABLE 10. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY MATERNAL PRENATAL CARE, HAWAII, 1986-2003

DIAGNOSIS	NONE		YES - EVER		YES - 1ST*		YES - 2ND*		YES - 3RD*	
	2,429		315,474		255,222		48,913		11,339	
	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
BRAIN & NERVOUS SYSTEM	**40**	**164.7**	**1357**	**43.0**	**811**	**31.8**	**247**	**50.5**	**58**	**51.2**
Neural tube defects	2	8.2	325	10.3	176	6.9	38	7.8	9	7.9
Anencephaly	2	8.2	118	3.7	56	2.2	14	2.9	0	0.0
Spina bifida	0	0.0	149	4.7	91	3.6	17	3.5	7	6.2
Encephalocele	0	0.0	67	2.1	35	1.4	7	1.4	2	1.8
Microcephaly	16	65.9	321	10.2	193	7.6	79	16.2	18	15.9
Holoprosencephaly	1	4.1	41	1.3	18	0.7	10	2.0	3	2.6
Hydrocephaly (w/o spina bifida)	12	49.4	364	11.5	220	8.6	65	13.3	18	15.9
EYE, EAR, FACE, & NECK	**49**	**201.7**	**1734**	**55.0**	**1166**	**45.7**	**334**	**68.3**	**79**	**69.7**
EYE	16	65.9	662	21.0	454	17.8	123	25.1	38	33.5
Anophthalmia	1	4.1	12	0.4	7	0.3	3	0.6	1	0.9
Microphthalmia	3	12.4	100	3.2	71	2.8	16	3.3	7	6.2
Cataract	0	0.0	44	1.4	35	1.4	5	1.0	3	2.6
EAR	35	144.1	1219	38.6	816	32.0	245	50.1	54	47.6
Absent auditory canal/ear	2	8.2	35	1.1	26	1.0	3	0.6	4	3.5
Microtia	1	4.1	119	3.8	79	3.1	30	6.1	3	2.6
CARDIAC & CIRCULATORY	**159**	**654.6**	**6174**	**195.7**	**4358**	**170.8**	**1038**	**212.2**	**260**	**229.3**
Common truncus	0	0.0	24	0.8	20	0.8	1	0.2	0	0.0
Transposition of the great vessels	6	24.7	140	4.4	84	3.3	28	5.7	11	9.7
Tetralogy of Fallot	2	8.2	128	4.1	92	3.6	18	3.7	7	6.2
Single ventricle	2	8.2	26	0.8	15	0.6	8	1.6	1	0.9
Ventricular septal defect	35	144.1	1567	49.7	1104	43.3	260	53.2	70	61.7
Atrial septal defect	64	263.5	2245	71.2	1595	62.5	396	81.0	100	88.2
Endocardial cushion defect	3	12.4	77	2.4	52	2.0	11	2.2	2	1.8
Pulmonary valve atresia/stenosis	6	24.7	316	10.0	223	8.7	44	9.0	16	14.1
Tricuspid valve atresia/stenosis	1	4.1	55	1.7	35	1.4	10	2.0	6	5.3
Ebstein's anomaly	0	0.0	16	0.5	10	0.4	2	0.4	1	0.9

TABLE 10. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY MATERNAL PRENATAL CARE, HAWAII, 1986-2003

DIAGNOSIS	NONE		YES - EVER		YES - 1ST*		YES - 2ND*		YES - 3RD*	
	2,429		315,474		255,222		48,913		11,339	
	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Aortic valve atresia/stenosis	0	0.0	48	1.5	32	1.3	12	2.5	1	0.9
Mitral valve atresia/stenosis	3	12.4	34	1.1	25	1.0	6	1.2	2	1.8
Hypoplastic left heart syndrome	1	4.1	52	1.6	24	0.9	13	2.7	2	1.8
Dextrocardia	2	8.2	50	1.6	30	1.2	10	2.0	6	5.3
Patent ductus arteriosus**	34	140.0	2642	83.7	1939	76.0	436	89.1	118	104.1
Coarctation of the aorta	0	0.0	93	2.9	68	2.7	16	3.3	1	0.9
Aorta stenosis	0	0.0	2	0.1	1	0.0	0	0.0	0	0.0
Pulmonary artery atresia/stenosis	19	78.2	607	19.2	437	17.1	105	21.5	19	16.8
Total anomalous pulmonary venous return	1	4.1	31	1.0	23	0.9	5	1.0	3	2.6
Partial anomalous pulmonary venous	0	0.0	17	0.5	12	0.5	4	0.8	0	0.0
Persistent fetal circulation	6	24.7	189	6.0	119	4.7	47	9.6	11	9.7
RESPIRATORY	**17**	**70.0**	**876**	**27.8**	**588**	**23.0**	**171**	**35.0**	**36**	**31.7**
Choanal atresia/stenosis	2	8.2	45	1.4	33	1.3	10	2.0	0	0.0
Lung cysts	0	0.0	26	0.8	22	0.9	4	0.8	1	0.9
Hypoplastic lung	3	12.4	203	6.4	123	4.8	43	8.8	6	5.3
OROFACIAL & GASTROINTESTINAL	**37**	**152.3**	**2010**	**63.7**	**1380**	**54.1**	**350**	**71.6**	**92**	**81.1**
Oral clefts	12	49.4	661	21.0	464	18.2	112	22.9	27	23.8
Cleft palate (isolated)	6	24.7	242	7.7	172	6.7	45	9.2	9	7.9
Cleft lip (±cleft palate)	6	24.7	420	13.3	292	11.4	68	13.9	18	15.9
TE fistula/Esophageal atresia	0	0.0	71	2.3	51	2.0	11	2.2	3	2.6
Pyloric stenosis	1	4.1	261	8.3	181	7.1	46	9.4	6	5.3
Atresia/stenosis small intestine	2	8.2	94	3.0	64	2.5	11	2.2	1	0.9
Anal atresia/stenosis	1	4.1	169	5.4	122	4.8	29	5.9	6	5.3
Hirschsprung's disease	1	4.1	80	2.5	58	2.3	13	2.7	2	1.8
Malrotation	0	0.0	95	3.0	61	2.4	19	3.9	4	3.5
Biliary atresia	0	0.0	35	1.1	17	0.7	13	2.7	1	0.9
GENITAL & URINARY	**59**	**242.9**	**3657**	**115.9**	**2530**	**99.1**	**663**	**135.5**	**187**	**164.9**

TABLE 10. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY MATERNAL PRENATAL CARE, HAWAII, 1986-2003

DIAGNOSIS	NONE		YES - EVER		YES - 1ST*		YES - 2ND*		YES - 3RD*	
	2,429		315,474		255,222		48,913		11,339	
	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Ovarian cyst	0	0.0	18	0.6	11	0.4	3	0.6	1	0.9
Undescended testicle**	11	45.3	1112	35.2	747	29.3	249	50.9	54	47.6
Hypospadias/Epispadias/Chordee	12	49.4	1108	35.1	810	31.7	185	37.8	53	46.7
Ambiguous genitalia	1	4.1	14	0.4	8	0.3	2	0.4	2	1.8
Absent/hypoplastic kidney	3	12.4	163	5.2	97	3.8	22	4.5	11	9.7
Cystic kidney	1	4.1	149	4.7	100	3.9	23	4.7	10	8.8
Hydronephrosis	4	16.5	425	13.5	299	11.7	60	12.3	21	18.5
Exstrophy bladder	0	0.0	8	0.3	5	0.2	1	0.2	1	0.9
Posterior urethral valves	1	4.1	21	0.7	16	0.6	1	0.2	0	0.0
LIMB & MUSCULOSKELETAL	**81**	**333.5**	**4487**	**142.2**	**3099**	**121.4**	**748**	**152.9**	**195**	**172.0**
Scoliosis	1	4.1	87	2.8	48	1.9	18	3.7	0	0.0
Hip dislocation	3	12.4	351	11.1	267	10.5	54	11.0	13	11.5
Clubfoot	20	82.3	1083	34.3	719	28.2	176	36.0	61	53.8
Polydactyly	8	32.9	591	18.7	427	16.7	96	19.6	19	16.8
Syndactyly	5	20.6	279	8.8	194	7.6	45	9.2	14	12.3
Limb reduction deformity - upper	4	16.5	114	3.6	70	2.7	25	5.1	5	4.4
Limb reduction deformity - lower	0	0.0	50	1.6	34	1.3	8	1.6	2	1.8
Craniosynostosis	2	8.2	165	5.2	125	4.9	19	3.9	6	5.3
Crouzon's disease	0	0.0	2	0.1	1	0.0	0	0.0	1	0.9
Apert syndrome	0	0.0	11	0.3	9	0.4	2	0.4	0	0.0
Goldenhar syndrome	0	0.0	19	0.6	15	0.6	3	0.6	0	0.0
Spina bifida occulta	0	0.0	10	0.3	8	0.3	0	0.0	1	0.9
Dwarfism	1	4.1	95	3.0	65	2.5	13	2.7	5	4.4
Osteogenesis imperfecta	0	0.0	15	0.5	11	0.4	3	0.6	0	0.0
Diaphragmatic hernia	1	4.1	84	2.7	50	2.0	20	4.1	1	0.9
Abdominal wall defects	6	24.7	293	9.3	167	6.5	52	10.6	9	7.9
Omphalocele	0	0.0	94	3.0	43	1.7	17	3.5	2	1.8

TABLE 10. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY MATERNAL PRENATAL CARE, HAWAII, 1986-2003

DIAGNOSIS	NONE		YES - EVER		YES - 1ST*		YES - 2ND*		YES - 3RD*	
	2,429		315,474		255,222		48,913		11,339	
	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Gastroschisis	5	20.6	112	3.6	68	2.7	23	4.7	4	3.5
Prune-belly syndrome	0	0.0	9	0.3	7	0.3	1	0.2	0	0.0
Poland syndrome	0	0.0	5	0.2	5	0.2	0	0.0	0	0.0
SKIN & INTEGUMENT	**60**	**247.0**	**2062**	**65.4**	**1387**	**54.3**	**421**	**86.1**	**135**	**119.1**
CHROMOSOME & SYNDROME	**25**	**102.9**	**1811**	**57.4**	**1050**	**41.1**	**200**	**40.9**	**48**	**42.3**
Down syndrome (trisomy 21)	4	16.5	501	15.9	263	10.3	65	13.3	12	10.6
Patau syndrome (trisomy 13)	3	12.4	62	2.0	33	1.3	2	0.4	3	2.6
Edwards syndrome (trisomy 18)	1	4.1	157	5.0	61	2.4	11	2.2	5	4.4
Cri du Chat syndrome	0	0.0	7	0.2	3	0.1	1	0.2	1	0.9
Wolff-Hirschorn syndrome	0	0.0	8	0.3	5	0.2	1	0.2	1	0.9
Autosomal anomalies in normal infant	0	0.0	137	4.3	108	4.2	13	2.7	1	0.9
Triploidy	1	4.1	33	1.0	15	0.6	0	0.0	0	0.0
Turner syndrome (45,X)	3	12.4	128	4.1	67	2.6	6	1.2	0	0.0
Klinefelter syndrome (47,XXY)	0	0.0	51	1.6	34	1.3	2	0.4	0	0.0
Absent spleen/Asplenia	1	4.1	10	0.3	8	0.3	1	0.2	0	0.0
Accessory spleen	1	4.1	39	1.2	28	1.1	3	0.6	1	0.9
Situs inversus	1	4.1	36	1.1	21	0.8	6	1.2	3	2.6
Conjoined twins	0	0.0	8	0.3	0	0.0	0	0.0	2	1.8
Tuberous sclerosis	1	4.1	12	0.4	10	0.4	1	0.2	1	0.9
Amniotic band syndrome	1	4.1	74	2.3	41	1.6	14	2.9	3	2.6
DiGeorge syndrome	0	0.0	21	0.7	13	0.5	5	1.0	3	2.6
Pierre-Robin sequence	0	0.0	31	1.0	21	0.8	7	1.4	1	0.9
Werdnig-Hoffman syndrome	0	0.0	14	0.4	10	0.4	1	0.2	1	0.9
Wolff-Parkinson-White syndrome	0	0.0	18	0.6	13	0.5	4	0.8	0	0.0
NEOPLASMS	**7**	**28.8**	**339**	**10.7**	**214**	**8.4**	**39**	**8.0**	**6**	**5.3**
CONGENITAL INFECTION	**11**	**45.3**	**252**	**8.0**	**159**	**6.2**	**56**	**11.4**	**15**	**13.2**
MATERNAL SUBSTANCE ABUSE	**690**	**2840.7**	**2137**	**67.7**	**931**	**36.5**	**751**	**153.5**	**297**	**261.9**

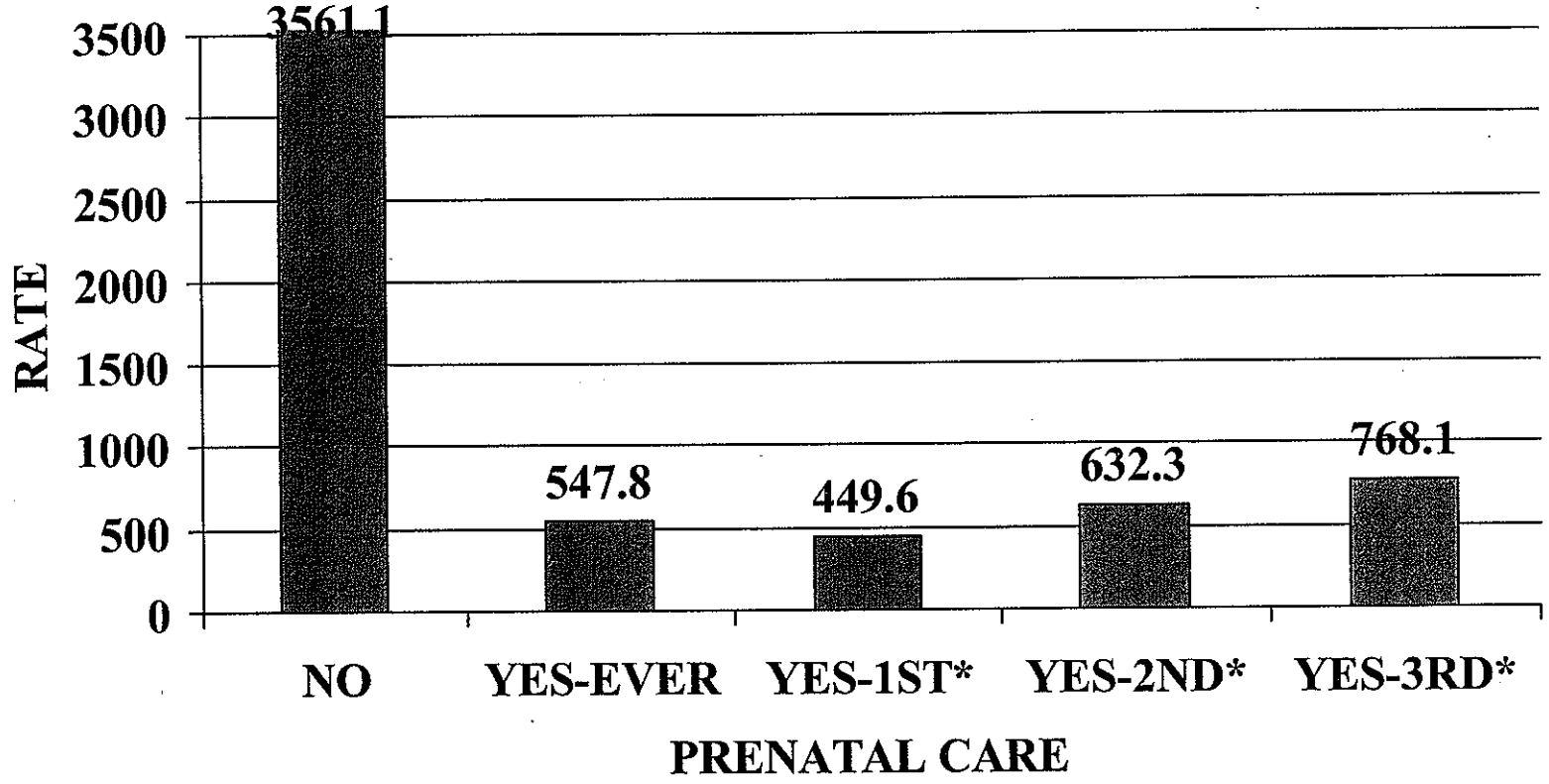
TABLE 10. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY MATERNAL PRENATAL CARE, HAWAII, 1986-2003

DIAGNOSIS	NONE		YES - EVER		YES - 1ST*		YES - 2ND*		YES - 3RD*	
	2,429		315,474		255,222		48,913		11,339	
	N	Rate	N	Rate	N	Rate	N	Rate	N	Rate
Fetal alcohol syndrome	13	53.5	75	2.4	31	1.2	25	5.1	14	12.3
Illicit drugs	685	2820.1	2092	66.3	911	35.7	737	150.7	288	254.0
Marijuana	147	605.2	763	24.2	389	15.2	251	51.3	76	67.0
Cocaine	137	564.0	428	13.6	189	7.4	152	31.1	52	45.9
Methamphetamine/Amphetamine	537	2210.8	1259	39.9	472	18.5	483	98.7	211	186.1
TOTAL	**865**	**3561.1**	**17283**	**547.8**	**11474**	**449.6**	**3093**	**632.3**	**871**	**768.1**

*Trimester

**Birth weight >2499 grams.

FIGURE 14. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY MATERNAL PRENATAL CARE, HAWAII, 1986-2003



*Trimester

TABLE 11. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY MATERNAL TOBACCO USE, HAWAII, 1988-2003*

DIAGNOSIS	NO		YES	
	266,783		27,305	
	N	Rate	N	Rate
BRAIN & NERVOUS SYSTEM	**953**	**35.7**	**281**	**102.9**
Neural tube defects	230	8.6	57	20.9
Anencephaly	75	2.8	25	9.2
Spina bifida	111	4.2	22	8.1
Encephalocele	50	1.9	12	4.4
Microcephaly	190	7.1	92	33.7
Holoprosencephaly	32	1.2	6	2.2
Hydrocephaly (w/o spina bifida)	258	9.7	72	26.4
EYE, EAR, FACE, & NECK	**1225**	**45.9**	**327**	**119.8**
EYE	469	17.6	133	48.7
Anophthalmia	9	0.3	3	1.1
Microphthalmia	72	2.7	23	8.4
Cataract	30	1.1	10	3.7
EAR	850	31.9	226	82.8
Absent auditory canal/ear	32	1.2	4	1.5
Microtia	82	3.1	20	7.3
CARDIAC & CIRCULATORY	**4490**	**168.3**	**1071**	**392.2**
Common truncus	20	0.7	1	0.4
Transposition of the great vessels	107	4.0	27	9.9
Tetralogy of Fallot	97	3.6	22	8.1
Single ventricle	20	0.7	4	1.5
Ventricular septal defect	1134	42.5	262	96.0
Atrial septal defect	1740	65.2	425	155.6
Endocardial cushion defect	50	1.9	20	7.3
Pulmonary valve atresia/stenosis	217	8.1	68	24.9
Tricuspid valve atresia/stenosis	37	1.4	14	5.1
Ebstein's anomaly	12	0.4	3	1.1
Aortic valve atresia/stenosis	28	1.0	11	4.0
Mitral valve atresia/stenosis	19	0.7	7	2.6
Hypoplastic left heart syndrome	35	1.3	10	3.7
Dextrocardia	31	1.2	13	4.8
Patent ductus arteriosus**	1960	73.5	428	156.7
Coarctation of the aorta	69	2.6	14	5.1
Aorta stenosis	2	0.1	0	0.0
Pulmonary artery atresia/stenosis	467	17.5	123	45.0
Total anomalous pulmonary venous return	26	1.0	3	1.1
Partial anomalous pulmonary venous	11	0.4	4	1.5
Persistent fetal circulation	106	4.0	22	8.1
RESPIRATORY	**608**	**22.8**	**169**	**61.9**

TABLE 11. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY MATERNAL TOBACCO USE, HAWAII, 1988-2003*

DIAGNOSIS	NO 266,783		YES 27,305	
	N	Rate	N	Rate
Choanal atresia/stenosis	36	1.3	6	2.2
Lung cysts	20	0.7	6	2.2
Hypoplastic lung	131	4.9	33	12.1
OROFACIAL & GASTROINTESTINAL	**1388**	**52.0**	**382**	**139.9**
Oral clefts	434	16.3	146	53.5
Cleft palate (isolated)	148	5.5	68	24.9
Cleft lip (±cleft palate)	286	10.7	78	28.6
TE fistula/Esophageal atresia	44	1.6	15	5.5
Pyloric stenosis	181	6.8	49	17.9
Atresia/stenosis small intestine	77	2.9	13	4.8
Anal atresia/stenosis	120	4.5	29	10.6
Hirschsprung's disease	58	2.2	6	2.2
Malrotation	70	2.6	15	5.5
Biliary atresia	26	1.0	4	1.5
GENITAL & URINARY	**2688**	**100.8**	**568**	**208.0**
Ovarian cyst	17	0.6	1	0.4
Undescended testicle**	774	29.0	179	65.6
Hypospadias/Epispadias/Chordee	870	32.6	132	48.3
Ambiguous genitalia	9	0.3	4	1.5
Absent/hypoplastic kidney	116	4.3	28	10.3
Cystic kidney	110	4.1	23	8.4
Hydronephrosis	336	12.6	63	23.1
Exstrophy bladder	7	0.3	1	0.4
Posterior urethral valves	16	0.6	2	0.7
LIMB & MUSCULOSKELETAL	**3145**	**117.9**	**771**	**282.4**
Scoliosis	60	2.2	13	4.8
Hip dislocation	265	9.9	42	15.4
Clubfoot	685	25.7	228	83.5
Polydactyly	432	16.2	102	37.4
Syndactyly	203	7.6	44	16.1
Limb reduction deformity - upper	75	2.8	29	10.6
Limb reduction deformity - lower	36	1.3	12	4.4
Craniosynostosis	119	4.5	22	8.1
Crouzon's disease	2	0.1	0	0.0
Apert syndrome	9	0.3	1	0.4
Goldenhar syndrome	11	0.4	5	1.8
Spina bifida occulta	7	0.3	3	1.1
Dwarfism	66	2.5	14	5.1
Osteogenesis imperfecta	12	0.4	3	1.1

TABLE 11. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY MATERNAL TOBACCO USE, HAWAII, 1988-2003*

DIAGNOSIS	NO 266,783		YES 27,305	
	N	Rate	N	Rate
Diaphragmatic hernia	59	2.2	14	5.1
Abdominal wall defects	219	8.2	50	18.3
Omphalocele	71	2.7	13	4.8
Gastroschisis	83	3.1	23	8.4
Prune-belly syndrome	8	0.3	1	0.4
Poland syndrome	4	0.1	1	0.4
SKIN & INTEGUMENT	**1404**	**52.6**	**454**	**166.3**
CHROMOSOME & SYNDROME	**1417**	**53.1**	**241**	**88.3**
Down syndrome (trisomy 21)	391	14.7	57	20.9
Patau syndrome (trisomy 13)	54	2.0	6	2.2
Edwards syndrome (trisomy 18)	132	4.9	14	5.1
Cri du Chat syndrome	7	0.3	0	0.0
Wolff-Hirschorn syndrome	6	0.2	2	0.7
Autosomal anomalies in normal infant	108	4.0	9	3.3
Triploidy	28	1.0	2	0.7
Turner syndrome (45,X)	99	3.7	16	5.9
Klinefelter syndrome (47,XXY)	39	1.5	6	2.2
Absent spleen/Asplenia	6	0.2	5	1.8
Accessory spleen	32	1.2	3	1.1
Situs inversus	26	1.0	7	2.6
Conjoined twins	10	0.4	2	0.7
Tuberous sclerosis	10	0.4	3	1.1
Amniotic band syndrome	43	1.6	22	8.1
DiGeorge syndrome	15	0.6	5	1.8
Pierre-Robin sequence	18	0.7	4	1.5
Werdnig-Hoffman syndrome	10	0.4	3	1.1
Wolff-Parkinson-White syndrome	14	0.5	3	1.1
NEOPLASMS	**259**	**9.7**	**44**	**16.1**
CONGENITAL INFECTION	**158**	**5.9**	**57**	**20.9**
MATERNAL SUBSTANCE ABUSE	**716**	**26.8**	**2040**	**747.1**
Fetal alcohol syndrome	19	0.7	58	21.2
Illicit drugs	704	26.4	2010	736.1
Marijuana	215	8.1	668	244.6
Cocaine	121	4.5	417	152.7
Methamphetamine/Amphetamine	448	16.8	1337	489.7
TOTAL	**11936**	**447.4**	**4151**	**1520.2**

*Denominators are not available prior to 1988.

**Birth weight >2499 grams.

FIGURE 15. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY MATERNAL TOBACCO USE, HAWAII, 1988-2003

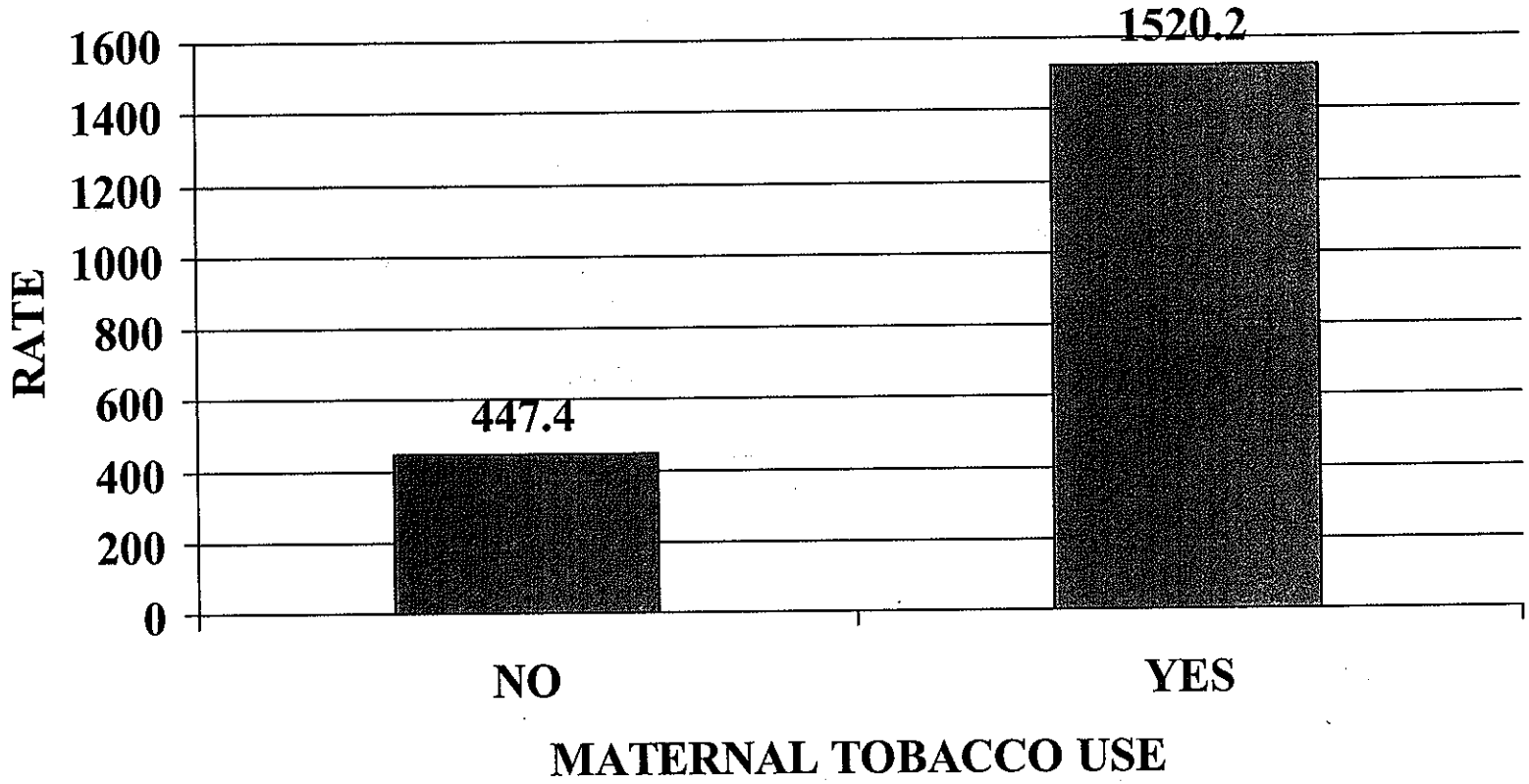


TABLE 12. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY MATERNAL ALCOHOL USE, HAWAII, 1988-2003*

DIAGNOSIS	NO		YES	
	290,327		3,440	
	N	Rate	N	Rate
BRAIN & NERVOUS SYSTEM	**1132**	**39.0**	**100**	**290.7**
Neural tube defects	262	9.0	20	58.1
Anencephaly	88	3.0	7	20.3
Spina bifida	122	4.2	11	32.0
Encephalocele	58	2.0	3	8.7
Microcephaly	245	8.4	39	113.4
Holoprosencephaly	35	1.2	3	8.7
Hydrocephaly (w/o spina bifida)	307	10.6	23	66.9
EYE, EAR, FACE, & NECK	**1415**	**48.7**	**148**	**430.2**
EYE	535	18.4	73	212.2
Anophthalmia	9	0.3	3	8.7
Microphthalmia	89	3.1	6	17.4
Cataract	39	1.3	1	2.9
EAR	987	34.0	98	284.9
Absent auditory canal/ear	33	1.1	3	8.7
Microtia	97	3.3	6	17.4
CARDIAC & CIRCULATORY	**5250**	**180.8**	**321**	**933.1**
Common truncus	19	0.7	2	5.8
Transposition of the great vessels	122	4.2	11	32.0
Tetralogy of Fallot	109	3.8	11	32.0
Single ventricle	23	0.8	1	2.9
Ventricular septal defect	1301	44.8	103	299.4
Atrial septal defect	2054	70.7	113	328.5
Endocardial cushion defect	64	2.2	6	17.4
Pulmonary valve atresia/stenosis	259	8.9	27	78.5
Tricuspid valve atresia/stenosis	45	1.5	6	17.4
Ebstein's anomaly	14	0.5	1	2.9
Aortic valve atresia/stenosis	35	1.2	4	11.6
Mitral valve atresia/stenosis	24	0.8	2	5.8
Hypoplastic left heart syndrome	42	1.4	3	8.7
Dextrocardia	40	1.4	3	8.7
Patent ductus arteriosus**	2289	78.8	101	293.6
Coarctation of the aorta	75	2.6	8	23.3
Aorta stenosis	2	0.1	0	0.0
Pulmonary artery atresia/stenosis	559	19.3	33	95.9
Total anomalous pulmonary venous return	28	1.0	1	2.9
Partial anomalous pulmonary venous	15	0.5	0	0.0
Persistent fetal circulation	124	4.3	4	11.6
RESPIRATORY	**714**	**24.6**	**63**	**183.1**

TABLE 12. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY MATERNAL ALCOHOL USE, HAWAII, 1988-2003*

DIAGNOSIS	NO 290,327		YES 3,440	
	N	Rate	N	Rate
Choanal atresia/stenosis	39	1.3	2	5.8
Lung cysts	25	0.9	1	2.9
Hypoplastic lung	159	5.5	5	14.5
OROFACIAL & GASTROINTESTINAL	**1622**	**55.9**	**158**	**459.3**
Oral clefts	543	18.7	44	127.9
Cleft palate (isolated)	202	7.0	18	52.3
Cleft lip (±cleft palate)	342	11.8	26	75.6
TE fistula/Esophageal atresia	56	1.9	3	8.7
Pyloric stenosis	212	7.3	18	52.3
Atresia/stenosis small intestine	84	2.9	6	17.4
Anal atresia/stenosis	136	4.7	12	34.9
Hirschsprung's disease	61	2.1	2	5.8
Malrotation	84	2.9	1	2.9
Biliary atresia	30	1.0	1	2.9
GENITAL & URINARY	**3076**	**105.9**	**186**	**540.7**
Ovarian cyst	17	0.6	1	2.9
Undescended testicle**	905	31.2	48	139.5
Hypospadias/Epispadias/Chordee	947	32.6	53	154.1
Ambiguous genitalia	13	0.4	0	0.0
Absent/hypoplastic kidney	133	4.6	10	29.1
Cystic kidney	128	4.4	5	14.5
Hydronephrosis	380	13.1	21	61.0
Exstrophy bladder	8	0.3	0	0.0
Posterior urethral valves	18	0.6	0	0.0
LIMB & MUSCULOSKELETAL	**3608**	**124.3**	**322**	**936.0**
Scoliosis	66	2.3	6	17.4
Hip dislocation	286	9.9	21	61.0
Clubfoot	843	29.0	70	203.5
Polydactyly	498	17.2	38	110.5
Syndactyly	221	7.6	26	75.6
Limb reduction deformity - upper	92	3.2	12	34.9
Limb reduction deformity - lower	44	1.5	4	11.6
Craniosynostosis	131	4.5	9	26.2
Crouzon's disease	2	0.1	0	0.0
Apert syndrome	10	0.3	0	0.0
Goldenhar syndrome	15	0.5	2	5.8
Spina bifida occulta	10	0.3	0	0.0
Dwarfism	75	2.6	5	14.5
Osteogenesis imperfecta	15	0.5	0	0.0

TABLE 12. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY MATERNAL ALCOHOL USE, HAWAII, 1988-2003*

DIAGNOSIS	NO		YES	
	290,327		3,440	
	N	Rate	N	Rate
Diaphragmatic hernia	69	2.4	3	8.7
Abdominal wall defects	250	8.6	21	61.0
Omphalocele	77	2.7	7	20.3
Gastroschisis	100	3.4	8	23.3
Prune-belly syndrome	9	0.3	0	0.0
Poland syndrome	5	0.2	0	0.0
SKIN & INTEGUMENT	**1678**	**57.8**	**183**	**532.0**
CHROMOSOME & SYNDROME	**1532**	**52.8**	**127**	**369.2**
Down syndrome (trisomy 21)	411	14.2	38	110.5
Patau syndrome (trisomy 13)	58	2.0	4	11.6
Edwards syndrome (trisomy 18)	138	4.8	8	23.3
Cri du Chat syndrome	6	0.2	1	2.9
Wolff-Hirschorn syndrome	8	0.3	0	0.0
Autosomal anomalies in normal infant	109	3.8	8	23.3
Triploidy	27	0.9	3	8.7
Turner syndrome (45,X)	107	3.7	7	20.3
Klinefelter syndrome (47,XXY)	42	1.4	4	11.6
Absent spleen/Asplenia	11	0.4	0	0.0
Accessory spleen	32	1.1	2	5.8
Situs inversus	32	1.1	1	2.9
Conjoined twins	12	0.4	0	0.0
Tuberous sclerosis	13	0.4	0	0.0
Amniotic band syndrome	52	1.8	13	37.8
DiGeorge syndrome	20	0.7	0	0.0
Pierre-Robin sequence	22	0.8	1	2.9
Werdnig-Hoffman syndrome	13	0.4	0	0.0
Wolff-Parkinson-White syndrome	16	0.6	1	2.9
NEOPLASMS	**285**	**9.8**	**18**	**52.3**
CONGENITAL INFECTION	**204**	**7.0**	**11**	**32.0**
MATERNAL SUBSTANCE ABUSE	**2117**	**72.9**	**635**	**1845.9**
Fetal alcohol syndrome	5	0.2	74	215.1
Illicit drugs	2115	72.8	592	1720.9
Marijuana	624	21.5	260	755.8
Cocaine	333	11.5	200	581.4
Methamphetamine/Amphetamine	1484	51.1	299	869.2
TOTAL	**14799**	**509.7**	**1301**	**3782.0**

*Denominators are not available prior to 1988.

**Birth weight >2499 grams.

FIGURE 16. RATE OF BIRTH DEFECTS PER 10,000 BIRTHS BY MATERNAL ALCOHOL USE, HAWAII, 1988-2003

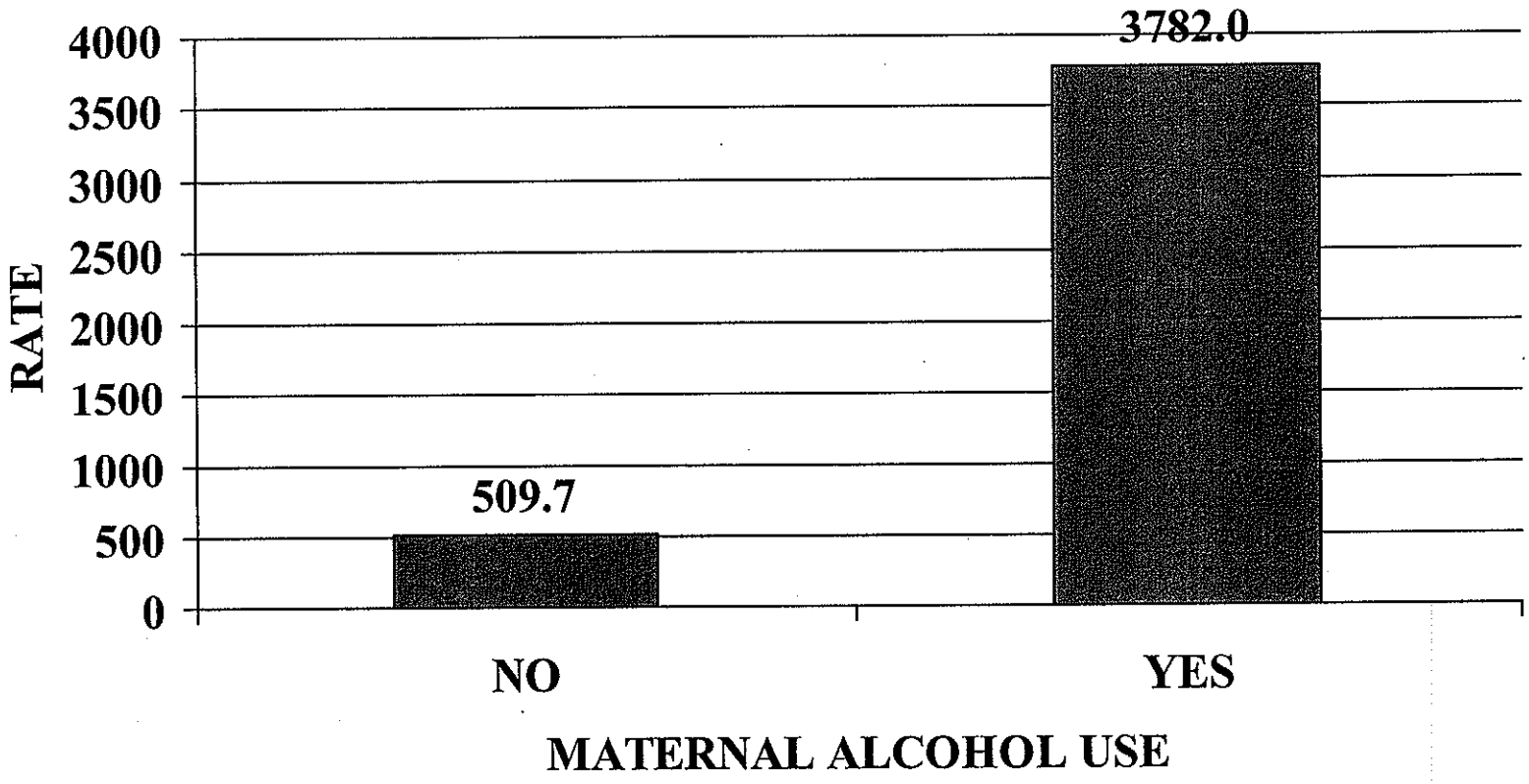


TABLE 13. PERCENTAGE OF BIRTH DEFECTS AFFECTED BY MATERNAL ILLICIT SUBSTANCE ABUSE, HAWAII, 1986-2003

DIAGNOSIS	N	%
BRAIN & NERVOUS SYSTEM	**78**	**5.5**
Neural tube defects	5	1.5
Anencephaly	1	0.8
Spina bifida	0	0.0
Encephalocele	4	5.9
Microcephaly	27	7.8
Holoprosencephaly	3	7.1
Hydrocephaly (w/o spina bifida)	19	4.9
EYE, EAR, FACE, & NECK	**105**	**5.8**
EYE	42	6.1
Anophthalmia	1	7.7
Microphthalmia	10	9.6
Cataract	1	2.3
EAR	72	5.7
Absent auditory canal/ear	2	5.4
Microtia	5	4.1
CARDIAC & CIRCULATORY	**347**	**5.4**
Common truncus	0	0.0
Transposition of the great vessels	9	6.1
Tetralogy of Fallot	7	5.3
Single ventricle	2	7.1
Ventricular septal defect	70	4.3
Atrial septal defect	135	5.7
Endocardial cushion defect	3	3.7
Pulmonary valve atresia/stenosis	13	3.9
Tricuspid valve atresia/stenosis	3	5.4
Ebstein's anomaly	1	5.6
Aortic valve atresia/stenosis	3	5.9
Mitral valve atresia/stenosis	2	5.4
Hypoplastic left heart syndrome	2	3.8
Dextrocardia	5	9.3
Patent ductus arteriosus*	119	4.4
Coarctation of the aorta	6	6.2
Aorta stenosis	0	0.0
Pulmonary artery atresia/stenosis	48	7.5
Total anomalous pulmonary venous return	0	0.0
Partial anomalous pulmonary venous	1	5.9
Persistent fetal circulation	6	3.0
RESPIRATORY	**47**	**5.2**
Choanal atresia/stenosis	1	2.0
Lung cysts	1	3.6

TABLE 13. PERCENTAGE OF BIRTH DEFECTS AFFECTED BY MATERNAL ILLICIT SUBSTANCE ABUSE, HAWAII, 1986-2003

DIAGNOSIS	N	%
Hypoplastic lung	8	3.8
OROFACIAL & GASTROINTESTINAL	**108**	**5.2**
Oral clefts	39	5.7
Cleft palate (isolated)	17	6.8
Cleft lip (±cleft palate)	22	5.1
TE fistula/Esophageal atresia	3	4.2
Pyloric stenosis	11	4.0
Atresia/stenosis small intestine	5	5.2
Anal atresia/stenosis	8	4.7
Hirschsprung's disease	0	0.0
Malrotation	2	2.1
Biliary atresia	1	2.8
GENITAL & URINARY	**136**	**3.6**
Ovarian cyst	0	0.0
Undescended testicle*	33	2.9
Hypospadias/Epispadias/Chordee	20	1.8
Ambiguous genitalia	0	0.0
Absent/hypoplastic kidney	5	3.0
Cystic kidney	5	3.2
Hydronephrosis	14	3.2
Exstrophy bladder	0	0.0
Posterior urethral valves	0	0.0
LIMB & MUSCULOSKELETAL	**214**	**4.6**
Scoliosis	2	2.2
Hip dislocation	8	2.2
Clubfoot	49	4.3
Polydactyly	25	4.2
Syndactyly	22	7.7
Limb reduction deformity - upper	12	10.1
Limb reduction deformity - lower	3	5.9
Craniosynostosis	1	0.6
Crouzon's disease	0	0.0
Apert syndrome	1	9.1
Goldenhar syndrome	2	10.5
Spina bifida occulta	0	0.0
Dwarfism	4	4.2
Osteogenesis imperfecta	1	6.7
Diaphragmatic hernia	3	3.5
Abdominal wall defects	12	4.0
Omphalocele	2	2.1
Gastroschisis	7	5.9

TABLE 13. PERCENTAGE OF BIRTH DEFECTS AFFECTED BY MATERNAL ILLICIT SUBSTANCE ABUSE, HAWAII, 1986-2003

DIAGNOSIS	N	%
Prune-belly syndrome	0	0.0
Poland syndrome	0	0.0
SKIN & INTEGUMENT	**242**	**11.2**
CHROMOSOME & SYNDROME	**50**	**2.6**
Down syndrome (trisomy 21)	10	1.9
Patau syndrome (trisomy 13)	2	3.0
Edwards syndrome (trisomy 18)	1	0.6
Cri du Chat syndrome	0	0.0
Wolff-Hirschorn syndrome	0	0.0
Autosomal anomalies in normal infant	2	1.5
Triploidy	0	0.0
Turner syndrome (45,X)	5	3.6
Klinefelter syndrome (47,XXY)	3	5.9
Absent spleen/Asplenia	2	18.2
Accessory spleen	0	0.0
Situs inversus	4	10.8
Conjoined twins	0	0.0
Tuberous sclerosis	0	0.0
Amniotic band syndrome	9	11.7
DiGeorge syndrome	0	0.0
Pierre-Robin sequence	2	6.5
Werdnig-Hoffman syndrome	0	0.0
Wolff-Parkinson-White syndrome	1	5.3
NEOPLASMS	**10**	**2.8**
CONGENITAL INFECTION	**14**	**5.2**
Fetal alcohol syndrome	38	42.7
TOTAL	**706**	**4.4**

*birth weight >2499 grams.

TABLE 14. DISTRIBUTION OF BIRTH DEFECTS BY PREGNANCY OUTCOME, HAWAII, 1986-2003

DIAGNOSIS	LIVEBIRTHS		FETAL DEATHS <20 WEEKS		FETAL DEATHS ≥20 WEEKS		TERMINATIONS	
	N	%*	N	%*	N	%*	N	%*
BRAIN & NERVOUS SYSTEM	**1123**	**78.9**	**16**	**1.1**	**55**	**3.9**	**229**	**16.1**
Neural tube defects	159	47.7	7	2.1	20	6.0	147	44.1
Anencephaly	19	15.8	5	4.2	14	11.7	82	68.3
Spina bifida	111	72.1	0	0.0	4	2.6	39	25.3
Encephalocele	32	47.1	2	2.9	3	4.4	31	45.6
Microcephaly	338	97.7	0	0.0	1	0.3	6	1.7
Holoprosencephaly	27	64.3	1	2.4	2	4.8	12	28.6
Hydrocephaly (w/o spina bifida)	300	77.9	5	1.3	25	6.5	55	14.3
EYE, EAR, FACE, & NECK	**1683**	**93.6**	**12**	**0.7**	**40**	**2.2**	**63**	**3.5**
EYE	667	97.2	0	0.0	8	1.2	11	1.6
Anophthalmia	7	53.8	0	0.0	3	23.1	3	23.1
Microphthalmia	101	97.1	0	0.0	2	1.9	1	1.0
Cataract	44	100.0	0	0.0	0	0.0	0	0.0
EAR	1192	94.3	8	0.6	29	2.3	35	2.8
Absent auditory canal/ear	37	100.0	0	0.0	0	0.0	0	0.0
Microtia	117	96.7	0	0.0	2	1.7	2	1.7
CARDIAC & CIRCULATORY	**6288**	**97.3**	**5**	**0.1**	**66**	**1.0**	**88**	**1.4**
Common truncus	19	79.2	0	0.0	1	4.2	4	16.7
Transposition of the great vessels	138	93.9	0	0.0	3	2.0	6	4.1
Tetralogy of Fallot	125	95.4	0	0.0	1	0.8	5	3.8
Single ventricle	25	89.3	0	0.0	1	3.6	2	7.1
Ventricular septal defect	1583	97.3	0	0.0	7	0.4	35	2.2
Atrial septal defect	2320	97.8	2	0.1	29	1.2	11	0.5
Endocardial cushion defect	72	88.9	0	0.0	4	4.9	5	6.2
Pulmonary valve atresia/stenosis	324	97.9	0	0.0	2	0.6	4	1.2
Tricuspid valve atresia/stenosis	54	96.4	0	0.0	0	0.0	2	3.6
Ebstein's anomaly	17	94.4	0	0.0	0	0.0	1	5.6
Aortic valve atresia/stenosis	50	98.0	0	0.0	1	2.0	0	0.0
Mitral valve atresia/stenosis	37	100.0	0	0.0	0	0.0	0	0.0
Hypoplastic left heart syndrome	42	79.2	0	0.0	2	3.8	9	17.0
Dextrocardia	51	94.4	0	0.0	0	0.0	2	3.7
Patent ductus arteriosus**	2697	99.8	0	0.0	6	0.2	0	0.0
Coarctation of the aorta	95	97.9	0	0.0	2	2.1	0	0.0
Aorta stenosis	2	100.0	0	0.0	0	0.0	0	0.0
Pulmonary artery atresia/stenosis	637	99.2	1	0.2	0	0.0	3	0.5
Total anomalous pulmonary venous return	32	100.0	0	0.0	0	0.0	0	0.0
Partial anomalous pulmonary venous	16	94.1	0	0.0	0	0.0	1	5.9
Persistent fetal circulation	201	100.0	0	0.0	0	0.0	0	0.0
RESPIRATORY	**851**	**93.6**	**3**	**0.3**	**25**	**2.8**	**30**	**3.3**

TABLE 14. DISTRIBUTION OF BIRTH DEFECTS BY PREGNANCY OUTCOME, HAWAII, 1986-2003

DIAGNOSIS	LIVEBIRTHS		FETAL DEATHS <20 WEEKS		FETAL DEATHS ≥20 WEEKS		TERMINATIONS	
	N	%*	N	%*	N	%*	N	%*
Choanal atresia/stenosis	50	100.0	0	0.0	0	0.0	0	0.0
Lung cysts	26	92.9	0	0.0	1	3.6	1	3.6
Hypoplastic lung	182	87.1	1	0.5	13	6.2	13	6.2
OROFACIAL & GASTROINTESTINAL	**1973**	**94.8**	**10**	**0.5**	**44**	**2.1**	**55**	**2.6**
Oral clefts	621	91.3	6	0.9	19	2.8	34	5.0
Cleft palate (isolated)	232	93.2	3	1.2	7	2.8	7	2.8
Cleft lip (±cleft palate)	389	90.0	3	0.7	13	3.0	27	6.3
TE fistula/Esophageal atresia	67	94.4	0	0.0	2	2.8	2	2.8
Pyloric stenosis	271	99.6	0	0.0	0	0.0	1	0.4
Atresia/stenosis small intestine	89	92.7	0	0.0	4	4.2	3	3.1
Anal atresia/stenosis	159	93.0	3	1.8	5	2.9	4	2.3
Hirschsprung's disease	84	100.0	0	0.0	0	0.0	0	0.0
Malrotation	90	92.8	0	0.0	2	2.1	5	5.2
Biliary atresia	35	97.2	0	0.0	1	2.8	0	0.0
GENITAL & URINARY	**3668**	**96.8**	**11**	**0.3**	**39**	**1.0**	**65**	**1.7**
Ovarian cyst	18	100.0	0	0.0	0	0.0	0	0.0
Undescended testicle**	1136	99.8	0	0.0	2	0.2	0	0.0
Hypospadias/Epispadias/Chordee	1129	98.9	2	0.2	2	0.2	3	0.3
Ambiguous genitalia	9	60.0	2	13.3	3	20.0	1	6.7
Absent/hypoplastic kidney	134	80.2	1	0.6	10	6.0	22	13.2
Cystic kidney	135	87.7	1	0.6	4	2.6	13	8.4
Hydronephrosis	413	94.7	2	0.5	5	1.1	15	3.4
Exstrophy bladder	8	88.9	0	0.0	0	0.0	1	11.1
Posterior urethral valves	16	72.7	2	9.1	0	0.0	4	18.2
LIMB & MUSCULOSKELETAL	**4395**	**94.8**	**21**	**0.5**	**73**	**1.6**	**146**	**3.1**
Scoliosis	69	76.7	2	2.2	2	2.2	17	18.9
Hip dislocation	359	99.7	0	0.0	1	0.3	0	0.0
Clubfoot	1089	96.6	2	0.2	13	1.2	23	2.0
Polydactyly	582	96.7	1	0.2	6	1.0	13	2.2
Syndactyly	270	94.7	3	1.1	4	1.4	8	2.8
Limb reduction deformity - upper	102	85.7	3	2.5	6	5.0	8	6.7
Limb reduction deformity - lower	44	86.3	1	2.0	2	3.9	4	7.8
Craniosynostosis	175	100.0	0	0.0	0	0.0	0	0.0
Crouzon's disease	2	100.0	0	0.0	0	0.0	0	0.0
Apert syndrome	11	100.0	0	0.0	0	0.0	0	0.0
Goldenhar syndrome	19	100.0	0	0.0	0	0.0	0	0.0
Spina bifida occulta	11	91.7	1	8.3	0	0.0	0	0.0
Dwarfism	83	86.5	0	0.0	4	4.2	9	9.4
Osteogenesis imperfecta	14	93.3	0	0.0	0	0.0	1	6.7

TABLE 14. DISTRIBUTION OF BIRTH DEFECTS BY PREGNANCY OUTCOME, HAWAII, 1986-2003

DIAGNOSIS	LIVEBIRTHS		FETAL DEATHS <20 WEEKS		FETAL DEATHS ≥20 WEEKS		TERMINATIONS	
	N	%*	N	%*	N	%*	N	%*
Diaphragmatic hernia	73	85.9	0	0.0	3	3.5	9	10.6
Abdominal wall defects	234	77.7	9	3.0	16	5.3	42	14.0
Omphalocele	56	58.9	2	2.1	10	10.5	27	28.4
Gastroschisis	102	86.4	4	3.4	4	3.4	8	6.8
Prune-belly syndrome	8	88.9	1	11.1	0	0.0	0	0.0
Poland syndrome	5	100.0	0	0.0	0	0.0	0	0.0
SKIN & INTEGUMENT	**2138**	**99.1**	**0**	**0.0**	**6**	**0.3**	**12**	**0.6**
CHROMOSOME & SYNDROME	**1144**	**59.4**	**271**	**14.1**	**97**	**5.0**	**411**	**21.4**
Down syndrome (trisomy 21)	302	58.1	31	6.0	8	1.5	177	34.0
Patau syndrome (trisomy 13)	25	37.3	11	16.4	6	9.0	25	37.3
Edwards syndrome (trisomy 18)	52	32.3	15	9.3	17	10.6	76	47.2
Cri du Chat syndrome	5	62.5	0	0.0	0	0.0	3	37.5
Wolff-Hirschorn syndrome	7	87.5	0	0.0	0	0.0	1	12.5
Autosomal anomalies in normal infant	127	92.7	2	1.5	4	2.9	4	2.9
Triploidy	0	0.0	34	77.3	5	11.4	5	11.4
Turner syndrome (45,X)	33	23.6	42	30.0	22	15.7	43	30.7
Klinefelter syndrome (47,XXY)	32	62.7	3	5.9	0	0.0	16	31.4
Absent spleen/Asplenia	7	63.6	0	0.0	3	27.3	1	9.1
Accessory spleen	29	70.7	0	0.0	9	22.0	3	7.3
Situs inversus	35	94.6	0	0.0	0	0.0	2	5.4
Conjoined twins	2	16.7	4	33.3	0	0.0	6	50.0
Tuberous sclerosis	13	100.0	0	0.0	0	0.0	0	0.0
Amniotic band syndrome	58	75.3	3	3.9	7	9.1	9	11.7
DiGeorge syndrome	21	100.0	0	0.0	0	0.0	0	0.0
Pierre-Robin sequence	31	100.0	0	0.0	0	0.0	0	0.0
Werdnig-Hoffman syndrome	11	78.6	0	0.0	1	7.1	2	14.3
Wolff-Parkinson-White syndrome	19	100.0	0	0.0	0	0.0	0	0.0
NEOPLASMS	**222**	**62.0**	**27**	**7.5**	**40**	**11.2**	**68**	**19.0**
CONGENITAL INFECTION	**248**	**92.9**	**1**	**0.4**	**14**	**5.2**	**4**	**1.5**
MATERNAL SUBSTANCE ABUSE	**2769**	**96.4**	**38**	**1.3**	**37**	**1.3**	**17**	**0.6**
Fetal alcohol syndrome	86	96.6	2	2.2	1	1.1	0	0.0
Illicit drugs	2719	96.4	38	1.3	36	1.3	17	0.6
Marijuana	887	96.0	13	1.4	15	1.6	5	0.5
Cocaine	550	95.5	13	2.3	8	1.4	3	0.5
Methamphetamine/Amphetamine	1771	97.0	17	0.9	20	1.1	11	0.6
TOTAL	**17209**	**92.7**	**352**	**1.9**	**279**	**1.5**	**685**	**3.7**

*Percentage of total.

**Birth weight >2499 grams.

FIGURE 17. DISTRIBUTION OF BIRTH DEFECTS BY PREGNANCY OUTCOME, HAWAII, 1986-2003

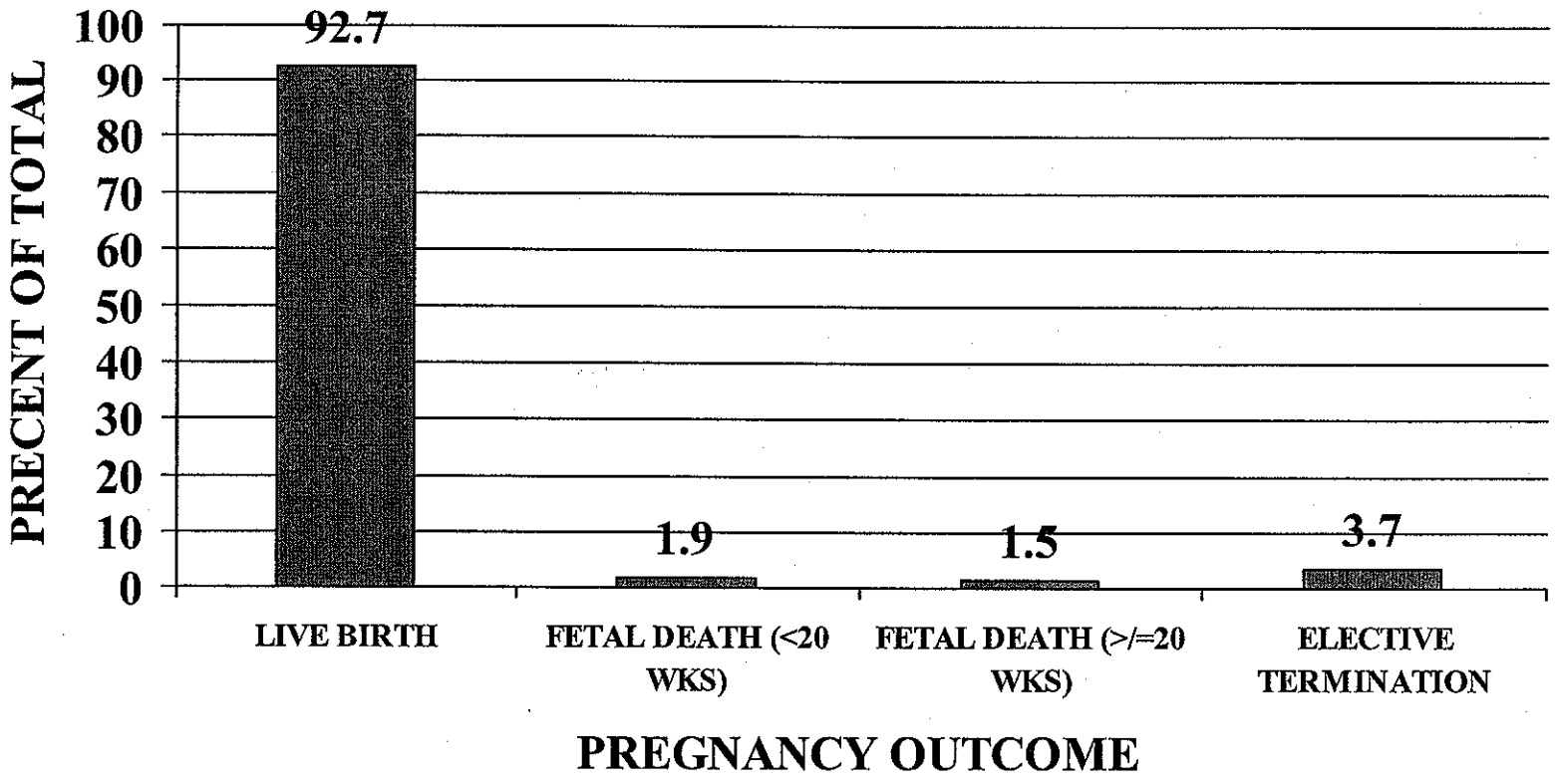


TABLE 15. MORTALITY RATES OF BIRTH DEFECTS, HAWAII, 1986-2002

DIAGNOSIS	N*	%**
BRAIN & NERVOUS SYSTEM	**175**	**16.3**
Neural tube defects	38	24.7
Anencephaly	19	100.0
Spina bifida	9	8.4
Encephalocele	13	41.9
Microcephaly	50	15.3
Holoprosencephaly	13	50.0
Hydrocephaly (w/o spina bifida)	54	18.7
EYE, EAR, FACE, & NECK	**224**	**14.0**
EYE	95	14.8
Anophthalmia	4	57.1
Microphthalmia	38	39.2
Cataract	3	7.3
EAR	185	16.3
Absent auditory canal/ear	4	11.4
Microtia	9	8.5
CARDIAC & CIRCULATORY	**531**	**9.0**
Common truncus	10	52.6
Transposition of the great vessels	30	23.1
Tetralogy of Fallot	18	15.0
Single ventricle	10	40.0
Ventricular septal defect	128	8.5
Atrial septal defect	269	12.7
Endocardial cushion defect	19	27.5
Pulmonary valve atresia/stenosis	32	10.3
Tricuspid valve atresia/stenosis	8	15.4
Ebstein's anomaly	2	13.3
Aortic valve atresia/stenosis	19	38.8
Mitral valve atresia/stenosis	23	62.2
Hypoplastic left heart syndrome	28	66.7
Dextrocardia	13	26.0
Patent ductus arteriosus***	156	6.3
Coarctation of the aorta	13	15.1
Aorta stenosis	1	50.0
Pulmonary artery atresia/stenosis	20	3.5
Total anomalous pulmonary venous return	6	19.4
Partial anomalous pulmonary venous	3	18.8
Persistent fetal circulation	38	19.8
RESPIRATORY	**199**	**24.1**
Choanal atresia/stenosis	4	8.2
Lung cysts	2	9.1
Hypoplastic lung	130	75.1

TABLE 15. MORTALITY RATES OF BIRTH DEFECTS, HAWAII, 1986-2002

DIAGNOSIS	N*	%**
OROFACIAL & GASTROINTESTINAL	**168**	**8.9**
Oral clefts	50	8.4
Cleft palate (isolated)	21	9.5
Cleft lip (±cleft palate)	29	7.8
TE fistula/Esophageal atresia	14	21.2
Pyloric stenosis	2	0.8
Atresia/stenosis small intestine	12	14.3
Anal atresia/stenosis	24	15.8
Hirschsprung's disease	9	11.1
Malrotation	13	14.8
Biliary atresia	4	12.1
GENITAL & URINARY	**210**	**6.0**
Ovarian cyst	0	0.0
Undescended testicle***	18	1.6
Hypospadias/Epispadias/Chordee	28	2.6
Ambiguous genitalia	2	22.2
Absent/hypoplastic kidney	46	37.1
Cystic kidney	32	24.8
Hydronephrosis	21	5.6
Exstrophy bladder	0	0.0
Posterior urethral valves	1	6.7
LIMB & MUSCULOSKELETAL	**280**	**6.6**
Scoliosis	19	28.8
Hip dislocation	15	4.3
Clubfoot	61	5.9
Polydactyly	20	3.6
Syndactyly	31	11.8
Limb reduction deformity - upper	27	27.0
Limb reduction deformity - lower	13	31.0
Craniosynostosis	10	5.7
Crouzon's disease	0	0.0
Apert syndrome	3	27.3
Goldenhar syndrome	2	10.5
Spina bifida occulta	2	18.2
Dwarfism	16	19.8
Osteogenesis imperfecta	6	42.9
Diaphragmatic hernia	30	42.9
Abdominal wall defects	30	13.4
Omphalocele	16	30.2
Gastroschisis	7	7.3
Prune-belly syndrome	3	37.5
Poland syndrome	0	0.0

TABLE 15. MORTALITY RATES OF BIRTH DEFECTS, HAWAII, 1986-2002

DIAGNOSIS	N*	%**
SKIN & INTEGUMENT	**108**	**5.3**
CHROMOSOME & SYNDROME	**182**	**16.5**
Down syndrome (trisomy 21)	22	7.7
Patau syndrome (trisomy 13)	20	83.3
Edwards syndrome (trisomy 18)	34	69.4
Cri du Chat syndrome	0	0.0
Wolff-Hirschorn syndrome	0	0.0
Autosomal anomalies in normal infant	4	3.2
Triploidy	0	
Turner syndrome (45,X)	2	6.1
Klinefelter syndrome (47,XXY)	0	0.0
Absent spleen/Asplenia	0	0.0
Accessory spleen	26	89.7
Situs inversus	4	11.8
Conjoined twins	2	100.0
Tuberous sclerosis	0	0.0
Amniotic band syndrome	17	29.3
DiGeorge syndrome	6	28.6
Pierre-Robin sequence	1	3.2
Werdnig-Hoffman syndrome	4	44.4
Wolff-Parkinson-White syndrome	1	5.6
NEOPLASMS	**21**	**9.9**
CONGENITAL INFECTION	**21**	**8.6**
MATERNAL SUBSTANCE ABUSE	**61**	**2.4**
Fetal alcohol syndrome	3	3.5
Illicit drugs	58	2.3
Marijuana	24	3.0
Cocaine	9	1.7
Methamphetamine/Amphetamine	26	1.6
TOTAL	**761**	**4.7**

*Number of livebirths which expired within one year of birth.

**Percentage of livebirths.

***Birth weight >2499 grams.

FIGURE 18. RATE PER 10,000 BIRTHS OF BIRTH DEFECTS BY PLURALITY, HAWAII, 1986-2003

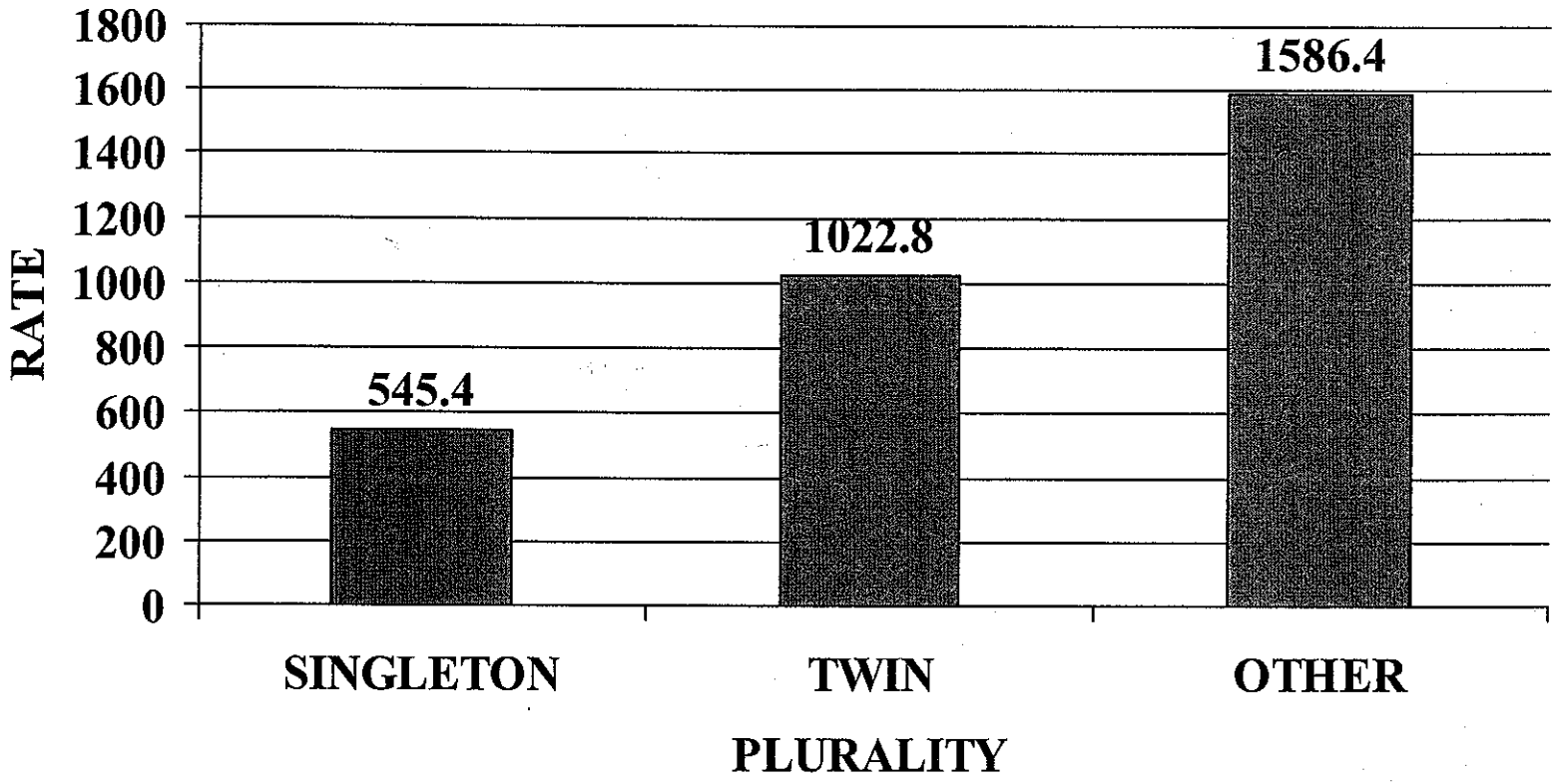


FIGURE 19. COMPARISON OF NATIVE HAWAIIAN VS NON-HAWAIIAN MOTHERS, HAWAII, 1986-2003

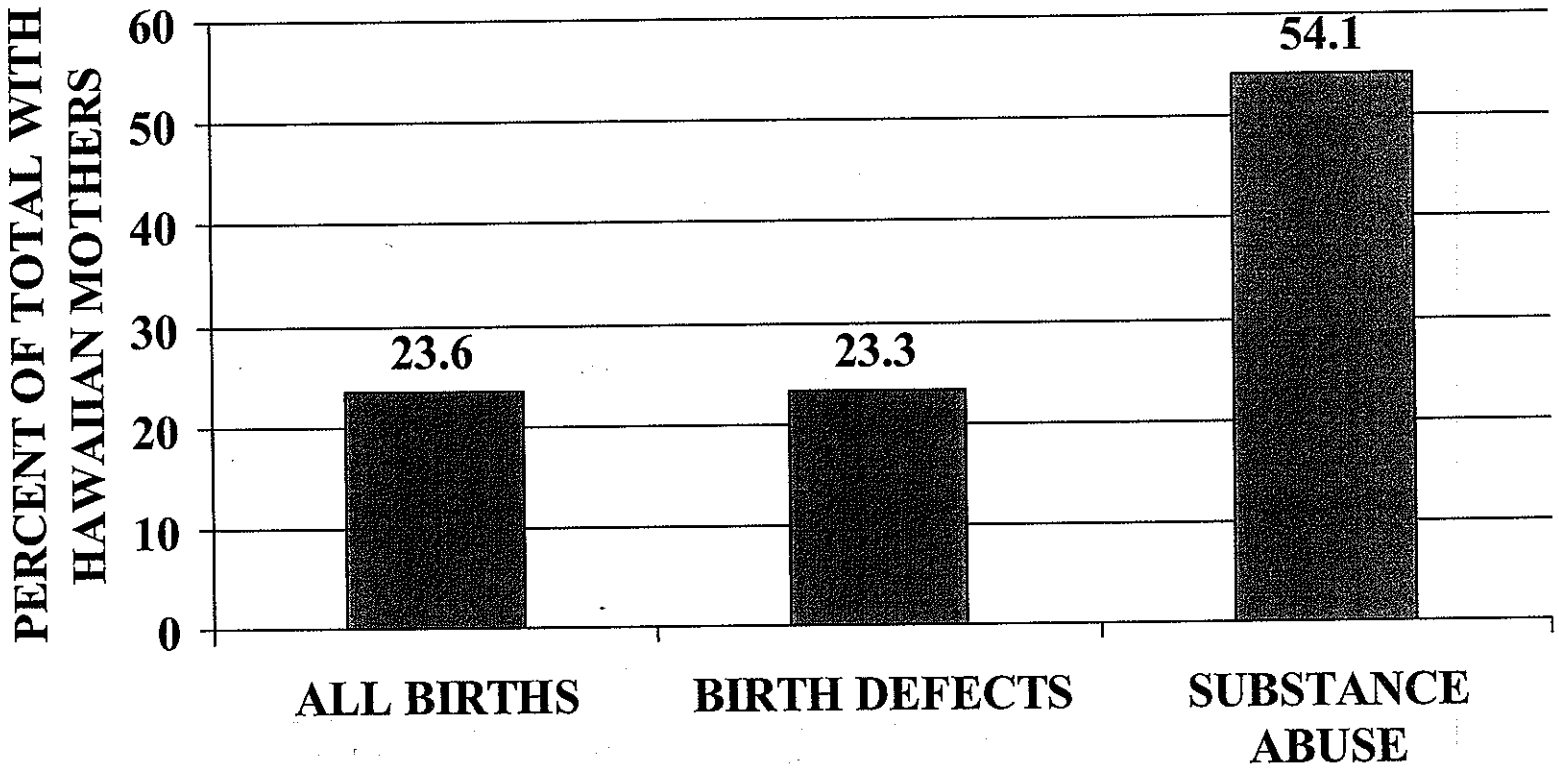


FIGURE 20. PROPORTION OF BIRTH DEFECT DIAGNOSES BY MAJOR ORGAN SYSTEM, HAWAII, 1986-2003

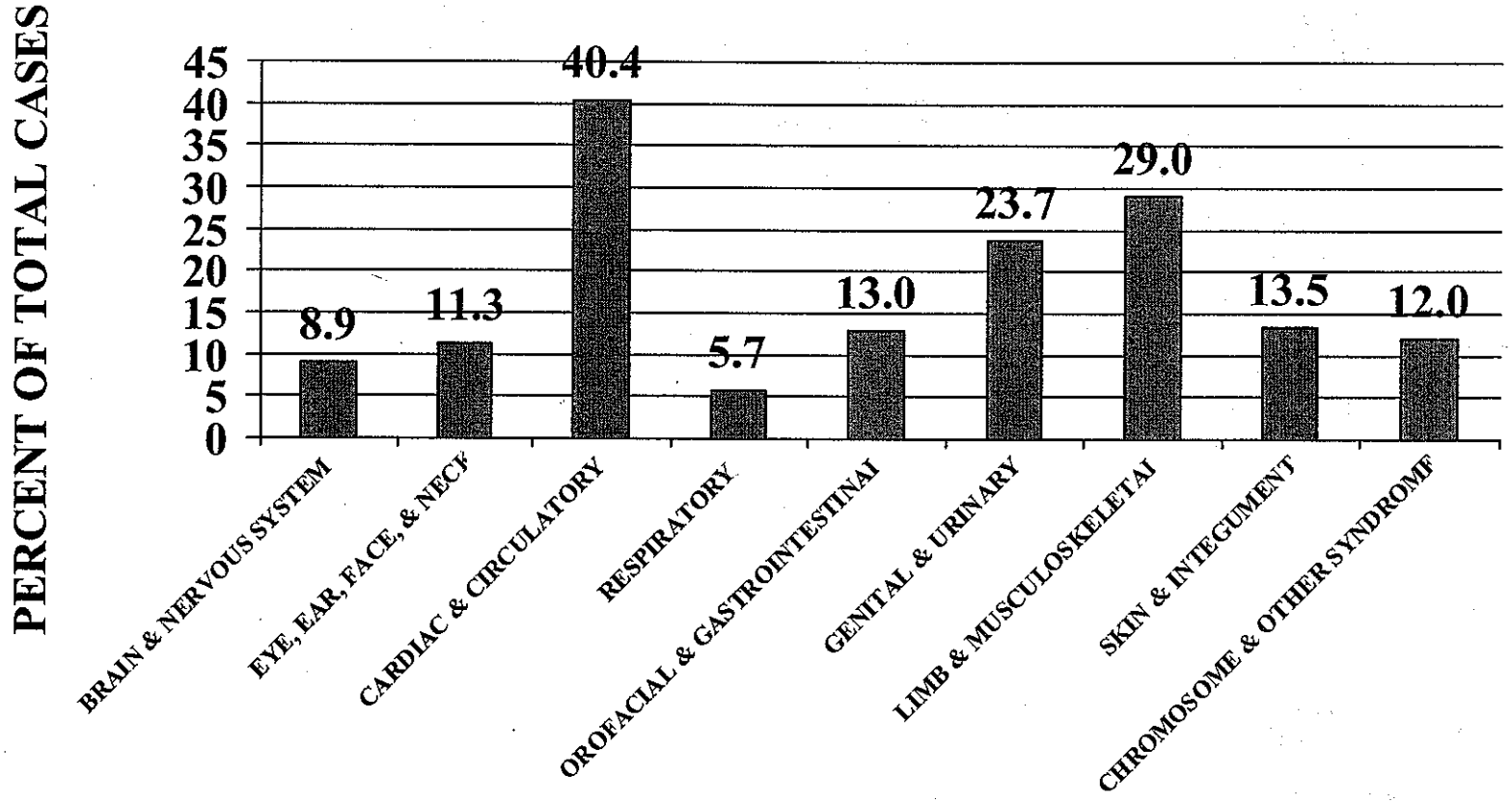


TABLE 16. PERCENTAGE OF BIRTH DEFECTS WITH ONE OR MORE OTHER DIAGNOSES, HAWAII, 1986-2003

DIAGNOSIS	N	%
BRAIN & NERVOUS SYSTEM	**1086**	**76.3**
Neural tube defects	200	60.1
Anencephaly	43	35.8
Spina bifida	111	72.1
Encephalocele	55	80.9
Microcephaly	294	85.0
Holoprosencephaly	40	95.2
Hydrocephaly (w/o spina bifida)	329	85.5
EYE, EAR, FACE, & NECK	**1617**	**89.9**
EYE	637	92.9
Anophthalmia	13	100.0
Microphthalmia	100	96.2
Cataract	25	56.8
EAR	1134	89.7
Absent auditory canal/ear	27	73.0
Microtia	77	63.6
CARDIAC & CIRCULATORY	**4772**	**73.9**
Common truncus	22	91.7
Transposition of the great vessels	144	98.0
Tetralogy of Fallot	123	93.9
Single ventricle	28	100.0
Ventricular septal defect	1168	71.8
Atrial septal defect	2285	96.4
Endocardial cushion defect	80	98.8
Pulmonary valve atresia/stenosis	304	91.8
Tricuspid valve atresia/stenosis	56	100.0
Ebstein's anomaly	17	94.4
Aortic valve atresia/stenosis	49	96.1
Mitral valve atresia/stenosis	37	100.0
Hypoplastic left heart syndrome	48	90.6
Dextrocardia	54	100.0
Patent ductus arteriosus*	1916	70.9
Coarctation of the aorta	88	90.7
Aorta stenosis	2	100.0
Pulmonary artery atresia/stenosis	504	78.5
Total anomalous pulmonary venous return	30	93.8
Partial anomalous pulmonary venous	16	94.1
Persistent fetal circulation	175	87.1
RESPIRATORY	**823**	**90.5**
Choanal atresia/stenosis	35	70.0
Lung cysts	16	57.1

TABLE 16. PERCENTAGE OF BIRTH DEFECTS WITH ONE OR MORE OTHER DIAGNOSES, HAWAII, 1986-2003

DIAGNOSIS	N	%
Hypoplastic lung	202	96.7
OROFACIAL & GASTROINTESTINAL	**1443**	**69.3**
Oral clefts	398	58.5
Cleft palate (isolated)	168	67.5
Cleft lip (±cleft palate)	231	53.5
TE fistula/Esophageal atresia	60	84.5
Pyloric stenosis	80	29.4
Atresia/stenosis small intestine	85	88.5
Anal atresia/stenosis	151	88.3
Hirschsprung's disease	43	51.2
Malrotation	92	94.8
Biliary atresia	24	66.7
GENITAL & URINARY	**2081**	**54.9**
Ovarian cyst	12	66.7
Undescended testicle*	412	36.2
Hypospadias/Epispadias/Chordee	519	45.5
Ambiguous genitalia	14	93.3
Absent/hypoplastic kidney	146	87.4
Cystic kidney	116	75.3
Hydronephrosis	332	76.1
Exstrophy bladder	9	100.0
Posterior urethral valves	21	95.5
LIMB & MUSCULOSKELETAL	**2882**	**62.1**
Scoliosis	88	97.8
Hip dislocation	162	45.0
Clubfoot	584	51.8
Polydactyly	277	46.0
Syndactyly	221	77.5
Limb reduction deformity - upper	107	89.9
Limb reduction deformity - lower	50	98.0
Craniosynostosis	100	57.1
Crouzon's disease	2	100.0
Apert syndrome	10	90.9
Goldenhar syndrome	19	100.0
Spina bifida occulta	10	83.3
Dwarfism	83	86.5
Osteogenesis imperfecta	12	80.0
Diaphragmatic hernia	76	89.4
Abdominal wall defects	210	69.8
Omphalocele	86	90.5
Gastroschisis	62	52.5

TABLE 16. PERCENTAGE OF BIRTH DEFECTS WITH ONE OR MORE OTHER DIAGNOSES, HAWAII, 1986-2003

DIAGNOSIS	N	%
Prune-belly syndrome	9	100.0
Poland syndrome	5	100.0
SKIN & INTEGUMENT	**2113**	**97.9**
CHROMOSOME & SYNDROME	**1203**	**62.5**
Down syndrome (trisomy 21)	345	66.3
Patau syndrome (trisomy 13)	52	77.6
Edwards syndrome (trisomy 18)	113	70.2
Cri du Chat syndrome	3	37.5
Wolff-Hirschorn syndrome	8	100.0
Autosomal anomalies in normal infant	42	30.7
Triploidy	6	13.6
Turner syndrome (45,X)	85	60.7
Klinefelter syndrome (47,XXY)	19	37.3
Absent spleen/Asplenia	11	100.0
Accessory spleen	40	97.6
Situs inversus	33	89.2
Conjoined twins	3	25.0
Tuberous sclerosis	9	69.2
Amniotic band syndrome	75	97.4
DiGeorge syndrome	21	100.0
Pierre-Robin sequence	30	96.8
Werdnig-Hoffman syndrome	9	64.3
Wolff-Parkinson-White syndrome	11	57.9
NEOPLASMS	**240**	**67.0**
CONGENITAL INFECTION	**187**	**70.0**
MATERNAL SUBSTANCE ABUSE	**1323**	**46.1**
Fetal alcohol syndrome	81	91.0
Illicit drugs	1280	45.4
Marijuana	649	70.2
Cocaine	290	50.3
Methamphetamine/Amphetamine	826	45.2
TOTAL	**9366**	**50.5**

*birth weight >2499 grams.

FIGURE 21. DISTRIBUTION OF BIRTH DEFECT CASES BY NUMBER OF DIAGNOSES, HAWAII, 1986-2003

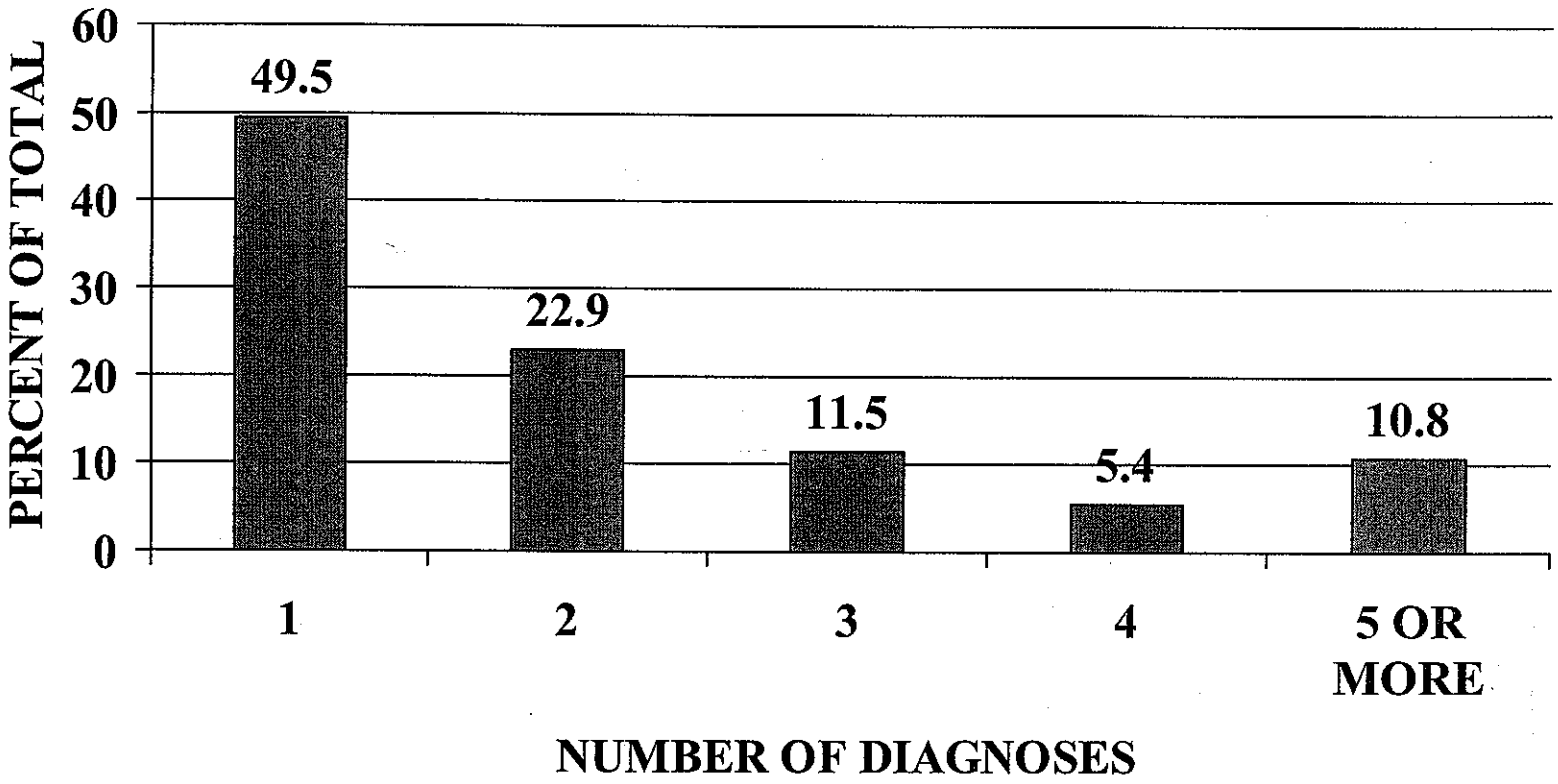
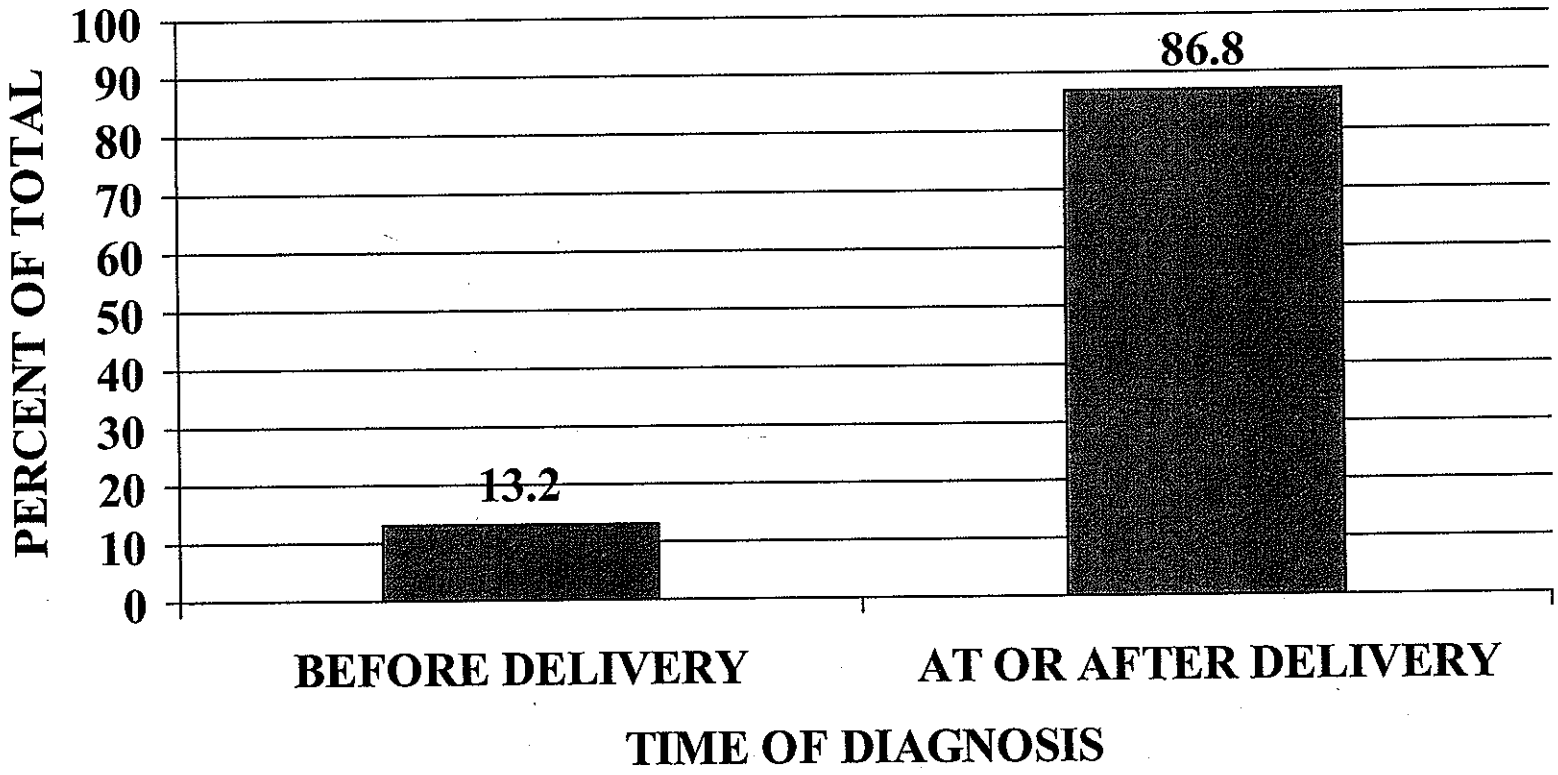


FIGURE 22. DISTRIBUTION OF BIRTH DEFECT CASES BY TIME OF DIAGNOSIS, HAWAII, 1986-2002



APPENDIX A. 34 PARTICIPATING HEALTH CARE FACILITIES

NAME	ISLAND
Castle Medical Center	Oahu
Fetal Diagnostic Institute of the Pacific	Oahu
Hana Medical Center	Maui
Hilo Medical Center	Hawaii
Hale Ho'ola Hamakua Hospital (formerly Honoka'a)*	Hawaii
Kahuku Hospital	Oahu
Kaiser-Permanente Medical Care	Oahu
Kapiolani Medical Center	Oahu
KMC Fetal Diagnostic Center	Oahu
KMC Genetics Section**	Oahu
KMC Molecular and Clinical Cytogenetics Laboratory**	Oahu
Kau Hospital	Hawaii
Kauai Veterans Memorial Hosp	Kauai
Kohala Hospital	Hawaii
Kona Community Hospital	Hawaii
Kula Hospital*	Maui
Lanai Community Hospital	Lanai
Maui Memorial Hospital	Maui
Medical Genetics Services**	Oahu
Mid-Pacific Genetics**	Oahu
Molokai General Hospital	Molokai
North Hawaii Community Hospital	Hawaii
Pali Momi Medical Center	Oahu
Queens Medical Center	Oahu
QMC Comprehensive Genetics Center	Oahu
QMC Genetics Laboratory	Oahu
Shriner's Hospital for Crippled 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

APPENDIX B. ICD-9 CODES OF INTEREST TO THE HBDP

090.0-090.2
155.0
159.8
162.8
171.8
186.0
189.0
190.5
191.0
194.0
202.3
208.0-208.9
214.0-214.9
216.0-216.9
228.0-228.1
237.7
238.0-238.9
239.2
255.2
257.8
279.11
335.0
426.7
427.9
524.0-524.1
551.1-551.3
553.2
630-635.9
637.0-637.9
655.0-655.9
656.4
740.0-759.9
760.70-760.79
768.0-768.1
771.0-771.2
774.4
779.9

APPENDIX C. BRITISH PEDIATRIC ASSOCIATION CODES FOR SPECIFIC DIAGNOSES IN THE HBDP

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 (birth weight>2499gm)
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
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 (birth weight>2499gm)
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	755.000-755.099
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
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
PIERRE-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

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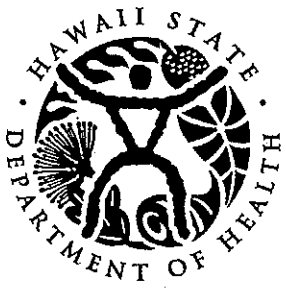

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