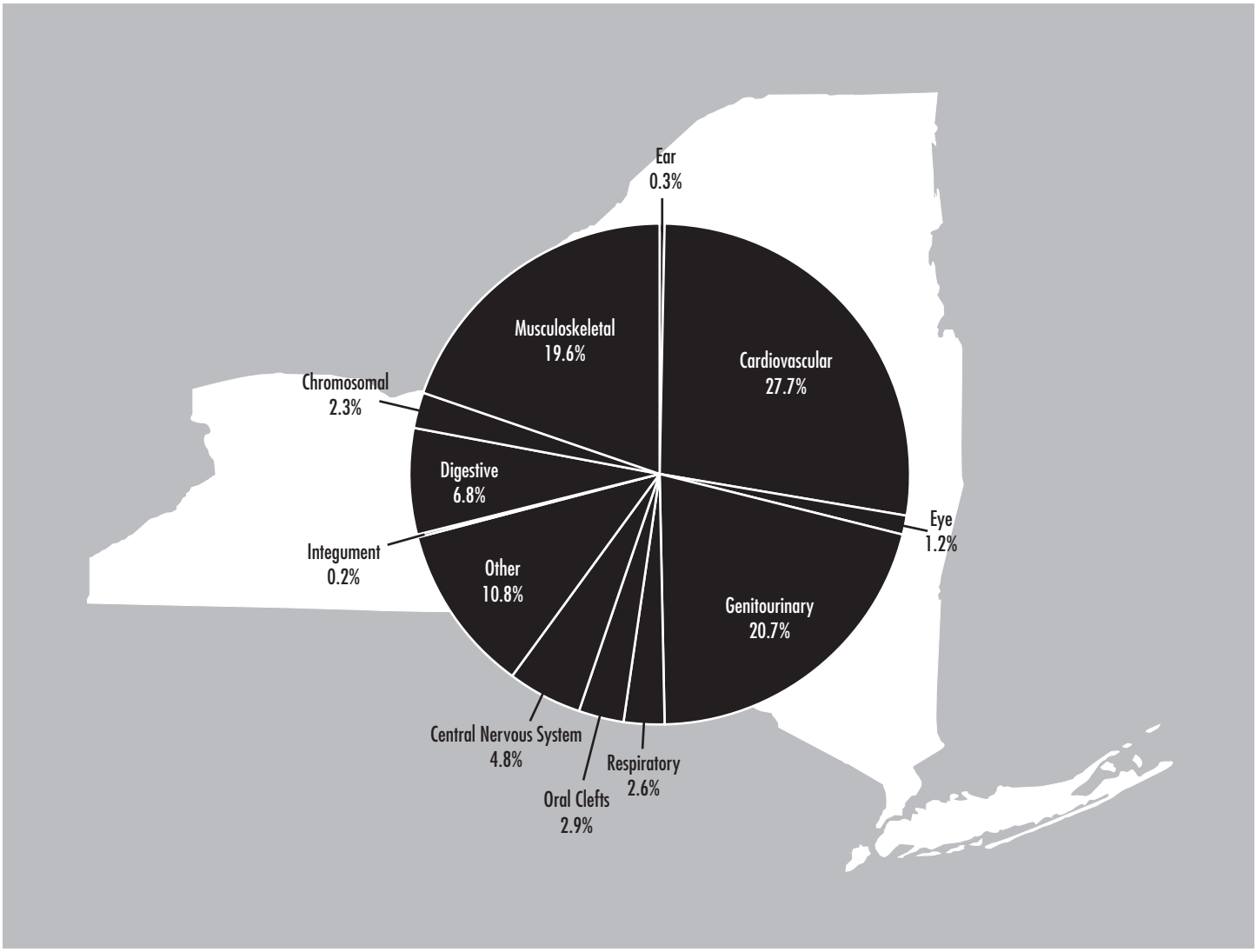
New York State Department of Health

Congenital Malformations Registry Summary Report



Statistical Summary of Children Born in 1996 and 1997 and Diagnosed Through 1999

New York State Department of Health

Congenital Malformations Registry Summary Report

Statistical Summary of Children Born in 1996-1997 and Diagnosed Through 1999

Additional and related information is also available from the New York State Department of Health Web site on the Internet: http://www.health.state.ny.us

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New York State Department of Health

Congenital Malformations Registry

Summary Report

Statistical Summary of Children Born in 1996-1997
and Diagnosed Through 1999

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Summary

The Congenital Malformations Registry Summary Report presents rates of congenital malformations occurring among the 520,587 children who were born alive to New York residents in 1996 and 1997. The children reported with a major congenital malformation represent 4.1% of live births. Males had a higher rate of major congenital malformations than females (4.9% versus 3.3%), and black children had a higher major malformation rate than white children (4.8% versus 4.0%). This information is provided through mandated reporting by hospitals and physicians.

Demographic characteristics of those children reported to the Congenital Malformations Registry (CMR), number of malformations and age at diagnosis are included in the report. Other sections present the distribution of anomalies by organ system; rates for selected malformations by race and sex and the most common malformations for each county in the state are also included.

This is the thirteenth report from the CMR. Reports are also available by request for the 1983 to 1995 birth cohorts. This report and the reports for 1994 and 1995 are also available on the Department of Health website. The statistics in this report are **not** comparable to reports before 1992. In 1992, the CMR began to use a new coding system that allows for greater detail in coding. For previous years, ICD-9 codes were used. Information from birth certificates was used to supplement or correct reported data. Birth certificate matching also helps eliminate duplicate cases reported under different names and nonresident births. Reports produced for 1989 to 1991 did not use birth certificate matching.

PROGRAM OVERVIEW

Background

Congenital malformations are the leading cause of infant mortality in the United States.[1] They are the fifth leading cause of years of potential life lost and a major cause of morbidity and mortality throughout childhood.[1,2] Twenty percent of infant deaths are attributed to congenital malformations,[2] a percentage that has increased over time.[1,2] Approximately 25% of pediatric hospital admissions and about one-third of the total number of pediatric hospital days are for congenital malformations of various types.[3] Little is known about the causes of congenital malformations. Twenty percent may be due to a combination of heredity and other factors; 7.5% may be due to single gene mutations; 6% to chromosome abnormalities; and 5% to maternal illnesses, such as diabetes, infections or anticonvulsant drugs.[4] Approximately 40% to 60% of congenital malformations are of unknown origin.[4,5]

Although radiation and rubella had been linked to birth defects, not until the thalidomide tragedy of the early 1960s was there a widespread interest in possible associations between congenital malformations and environmental agents. During the 1970s, interest continued to grow in birth defects and birth defects surveillance as a result of the growing recognition of the problems of toxic waste dumps such as Love Canal and accidents such as Three Mile Island and Seveso. In response, many states began to develop birth defects registries in order to have data for tracking trends in malformation rates.[6,7] A birth defects registry also makes it possible to respond to public concerns about possible excess occurrence of malformations with timely, objective investigations. A birth defects registry can provide cases for traditional epidemiologic studies of specific congential malformations and provide information for the planning, provision and evaluation of health services.[6,7]

New York State
Congenital Malformations Registry

The New York State Department of Health Congenital Malformations Registry (CMR) is one of the largest statewide, population-based birth defects registries in the nation. The concept of the Congenital Malformations Registry arose out of recognition of the environment as a potential etiologic factor in the occurrence of congenital malformations. Health studies during the Love Canal crisis in 1978 to 1983 confirmed the inadequacies of relying on birth certificates to monitor and evaluate birth defects.

New York's Congenital Malformations Registry was established by enactment of Part 22 of the State Sanitary Code in 1981. Reporting to the registry began in October 1982. Hospitals and physicians are required to report children under two years of age diagnosed with a malformation. The majority of reports are sent by hospitals, primarily from their medical records departments. A small number are sent by individual physicians to verify diagnosis initially suspected in the hospital but confirmed on an outpatient basis, and to clarify nonspecific diagnosis reported by hospitals.

The Congenital Malformations Registry receives case reports on children diagnosed up to two years of age who were born or reside in New York State with a congenital malformation, chromosomal anomaly or persistent metabolic defect. For purposes of this registry and report, a congenital malformation is defined as any structural, functional or biochemical abnormality, determined genetically or induced during gestation and not due to birthing events.

Case reports are received on forms (see Appendix 1) provided by the Department of Health (DOH). Pertinent fields are coded and the narrative description of the malformation is converted to a

code. The case report is matched to existing registry reports for possible duplicates. Data from reporting forms are entered on microcomputers and then transferred to the DOH mainframe for updating of the master files.

All information reported to the registry is held in strict confidence. Records and computer files are maintained in accordance with DOH regulations concerning data containing individual identifiers. Access to the data by anyone other than registry personnel is restricted and carefully monitored to ensure that confidentiality is maintained. Families of children reported to the registry are never contacted without prior consent of the DOH's Institutional Review Board and notification of the child's physician.

1996-1997 Report

This current report presents statistics for major anomalies only (see Appendix 2). This is in accordance with the practices of other state birth defects registries and allows comparison between New York State rates and rates in other states. Minor anomalies may cause problems in the determination of malformation rates because they are common and variably reported. They may not even be recorded in the medical chart.

The statistics in this report are **not** comparable to reports prior to 1992. The 1996-1997 report is based on birth certificate matched cases with resident live births from the vital records file used as the denominator. The available birth certificate fields are used to supplement or correct reported data. Birth certificate data are used to establish maternal residence at birth. Birth certificate matching helps eliminate duplicate cases reported under different names. Racial data are not comparable because race is defined by maternal race from the birth certificate. Using maternal race is a common practice among birth defects registries nationwide as the race of the father is poorly reported. In earlier years, race was defined by what was reported on the CMR form, which may differ from what is recorded on the birth certificate. In 1992, the registry began using a new coding system, the modified British Pediatric Association code (BPA). This coding scheme is used by a number of other congenital malformations registries and allows for greater specificity than does the ICD-9 system. Since 1992, the list of major malformations has been revised (see Appendix 4) changing the list of major malformations used in Sections I and II and the number of specific malformation prevalences in Section III.

CMR Birth Cohort reports are intended as a resource for programs providing primary, secondary and tertiary preventive health care and for public officials concerned with reducing overall mortality and morbidity. The first annual cohort included children born in 1983 and reported with a malformation diagnosed before their second birthday.[9] This report describes children born in 1996-1997 and diagnosed before their second birthday. Reports are also available for the 1984 through 1995 birth cohorts. Some reports and additional information are available through the DOH Web site at http://www.health.state.ny.us.

Limitations

Care should be taken in the use of these data. Virtually all reports are abstracted from inpatient hospital records, since malformations diagnosed on an outpatient basis are not well reported. Accurate hospital clinical recognition of malformations depends on clinical acumen and interest. This is particularly true of conditions more difficult to diagnose, such as fetal alcohol syndrome. Consequently, identification of malformations may vary by area and by time. The abstracting of records requires well-trained medical records professionals who are fastidious in their reporting of such findings. Areas with hospitals that provide higher levels of care may have more thorough diagnoses and, thus, apparently higher rates. Similarly, areas with hospitals that report cases more completely will also appear to have higher rates. In regions with low numbers of births, small variations in incidence may produce large statistical fluctuations.

New York State Population

Based on population projections from the 1990 census, the 1997 population of New York State was about 18.6 million; more than 40% of the population lived in New York City. An additional 23% of the population lived in the six counties closest to New York City. In 1996-1997, there were 520,587 resident live births reported to the state's vital registration, 22% to black mothers, and 19.8% to Hispanic mothers. In accordance with the practices of other state birth defects registries, the race of the child is based on race of the mother only. Nearly 46.5% of live births were to New York City residents.

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NOTES

Section I
Demographic Characteristics of Children Reported with Major Malformations

Introduction to Tables

These tables are based on children resident in New York State who were live born in 1996 and 1997 and reported to the registry with major malformations. Since a new coding system began to be used in 1992, the list of major malformations has been revised (see Appendix 4). Thus, the prevalence in this report are not comparable to reports prior to 1992.

The overall occurrences of major malformations for the two years are 4.1% to 4.2% of live births. Male children have a higher rate of major malformations than female children (4.8% to 4.9% versus 3.3% to 3.4%, Tables 1a, 1b). This difference is consistent within different racial groups. The rates for major malformations are somewhat higher for black than for white children (4.7% to 5.0% versus 4.0% to 4.1%). The major malformation rate among children with residence at birth in New York State excluding New York City was slightly higher than children with residence at birth in New York City (4.2% to 4.3% versus 4.0% to 4.1%). The smaller number of births in the "other" racial category makes these rates difficult to interpret.

About 77% of children reported with major malformations have only one major malformation (Tables 2a, 2b). Since most children had one major malformation, the race-sex patterns seen for all major malformations are similar to the patterns seen in children with a single major malformation (Tables 3a, 3b). All race-sex groups for children with multiple major malformations showed little variation (Tables 4a, 4b).

Section I - Table 1a
1996 Births – New York State Residents
Percent of Live Births With One Or More Major Malformations
Sex by Race and Residence

	Both Sexes			Males			Females		
Race and Residence	Infants	Total Births	%	Infants	Total Births	%	Infants	Total Births	%
New York State									
- All Races	11,020	263,611	4.2	6,664	135,084	4.9	4,356	128,527	3.4
- White	7,672	189,316	4.1	4,686	97,060	4.8	2,986	92,256	3.2
- Black	2,730	54,919	5.0	1,612	27,917	5.8	1,118	27,002	4.1
- Other	547	17,525	3.1	321	9,161	3.5	226	8,364	2.7
NYS Excluding NYC									
- All Races	6,000	140,661	4.3	3,717	72,131	5.2	2,283	68,530	3.3
- White	5,039	121,069	4.2	3,134	62,125	5.0	1,905	58,944	3.2
- Black	785	14,544	5.4	461	7,354	6.3	324	7,190	4.5
- Other	140	4,273	3.3	95	2,244	4.2	45	2,029	2.2
New York City									
- All Races	5,020	122,950	4.1	2,947	62,953	4.7	2,073	59,997	3.5
- White	2,633	68,247	3.9	1,552	34,935	4.4	1,081	33,312	3.2
- Black	1,945	40,375	4.8	1,151	20,563	5.6	794	19,812	4.0
- Other	407	13,252	3.1	226	6,917	3.3	181	6,335	2.9

Note: Total includes unknowns within each category, thus row and column figures may not sum to totals.

Section I - Table 1b
1997 Births – New York State Residents
Percent of Live Births With One Or More Major Malformations
Sex by Race and Residence

	Both Sexes			Males			Females		
Race and Residence	Infants	Total Births	%	Infants	Total Births	%	Infants	Total Births	%
New York State									
- All Races	10,457	256,976	4.1	6,324	131,353	4.8	4,128	125,619	3.3
- White	7,307	183,073	4.0	4,466	93,469	4.8	2,837	89,601	3.2
- Black	2,561	54,948	4.7	1,497	28,042	5.3	1,064	26,906	4.0
- Other	558	17,847	3.1	340	9,286	3.7	217	8,560	2.5
NYS Excluding NYC									
- All Races	5,749	138,074	4.2	3,556	70,594	5.0	2,188	67,476	3.2
- White	4,807	118,658	4.1	2,981	60,694	4.9	1,822	57,961	3.1
- Black	786	14,467	5.4	468	7,375	6.3	318	7,092	4.5
- Other	142	4,404	3.2	95	2,247	4.2	46	2,156	2.1
New York City									
- All Races	4,708	118,902	4.0	2,768	60,759	4.6	1,940	58,143	3.3
- White	2,500	64,415	3.9	1,485	32,775	4.5	1,015	31,640	3.2
- Black	1,755	40,481	4.3	1,029	20,667	5.0	746	19,814	3.8
- Other	416	13,443	3.1	245	7,039	3.5	171	6,404	2.7

Note: Total includes unknowns within each category, thus row and column figures may not sum to totals.

Section I – Table 2a
1996 Births – New York State Residents
Number of Major Malformations Per Child

Number of Malformations	Number of Children	Percent
1	8,493	77.1
2	1,577	14.3
3	482	4.4
4	221	2.0
5	102	0.9
6	50	0.5
7	35	0.3
8	17	0.2
9	14	0.1
10	12	0.1
11	8	0.1
12	6	0.1
>12	3	*
All Children	11,020	100.00

Less than 0.05%
Note: Total percent may not add to 100% due to rounding.

Section I – Table 2b
1997 Births – New York State Residents
Number of Major Malformations Per Child

Number of Malformations	Number of Children	Percent
1	8,104	77.5
2	1,505	14.4
3	455	4.4
4	180	1.7
5	88	0.8
6	55	0.5
7	32	0.3
8	18	0.2
9	8	0.1
10	7	0.1
11	2	*
12	2	*
>12	1	*
All Children	10,457	100.0

Less than 0.05%
Note: Total percent may not add to 100% due to rounding.

Section I - Table 3a
1996 Births – New York State Residents
Percent of Live Births With One Major Malformations
Sex by Race and Residence

	Both Sexes			Males			Females		
Race and Residence	Infants	Total Births	%	Infants	Total Births	%	Infants	Total Births	%
New York State									
- All Races	8,493	263,611	3.2	5,219	135,084	3.9	3,274	128,527	2.5
- White	5,881	189,316	3.1	3,663	97,060	3.8	2,218	92,256	2.4
- Black	2,134	54,919	3.9	1,279	27,917	4.6	855	27,002	3.2
- Other	421	17,525	2.4	242	9,161	2.6	179	8,364	2.1
NYS Excluding NYC									
- All Races	4,566	140,661	3.2	2,898	72,131	4.0	1,668	68,530	2.4
- White	3,835	121,069	3.2	2,443	62,125	3.9	1,392	58,944	2.4
- Black	598	14,544	4.1	363	7,354	4.9	235	7,190	3.3
- Other	104	4,273	2.4	70	2,244	3.1	34	2,029	1.7
New York City									
- All Races	3,927	122,950	3.2	2,321	62,953	3.7	1,606	59,997	2.7
- White	2,046	68,247	3.0	1,220	34,935	3.5	826	33,312	2.5
- Black	1,536	40,375	3.8	916	20,563	4.5	620	19,812	3.1
- Other	317	13,252	2.4	172	6,917	2.5	145	6,335	2.3

Note: Total includes unknowns within each category, thus row and column figures may not sum to totals.

Section I - Table 3b
1997 Births – New York State Residents
Percent of Live Births With One Major Malformations
Sex by Race and Residence

	Both Sexes			Males			Females		
Race and Residence	Infants	Total Births	%	Infants	Total Births	%	Infants	Total Births	%
New York State									
- All Races	8,104	256,976	3.2	4,954	131,353	3.8	3,150	125,619	2.5
- White	5,619	183,073	3.1	3,467	93,469	3.7	2,152	89,601	2.4
- Black	2,017	54,948	3.7	1,199	28,042	4.3	818	26,906	3.0
- Other	441	17,847	2.5	269	9,286	2.9	172	8,560	2.0
NYS Excluding NYC									
- All Races	4,413	138,074	3.2	2,753	70,594	3.9	1,660	67,476	2.5
- White	3,668	118,658	3.1	2,290	60,694	3.8	1,378	57,961	2.4
- Black	622	14,467	4.3	377	7,375	5.1	245	7,092	3.5
- Other	109	4,404	2.5	74	2,247	3.3	35	2,156	1.6
New York City									
- All Races	3,691	118,902	3.1	2,201	60,759	3.6	1,490	58,143	2.6
- White	1,951	64,415	3.0	1,177	32,775	3.6	774	31,640	2.4
- Black	1,395	40,481	3.4	822	20,667	4.0	573	19,814	2.9
- Other	332	13,443	2.5	195	7,039	2.8	137	6,404	2.1

Note: Total includes unknowns within each category, thus row and column figures may not sum to totals.

Section I - Table 4a
1996 Births – New York State Residents
Percent of Live Births With Two Or More Major Malformations
Sex by Race and Residence

	Both Sexes			Males			Females		
Race and Residence	Infants	Total Births	%	Infants	Total Births	%	Infants	Total Births	%
New York State									
- All Races	2,527	263,611	1.0	1,445	135,084	1.1	1,082	128,527	0.8
- White	1,791	189,316	1.0	1,023	97,060	1.1	768	92,256	0.8
- Black	596	54,919	1.1	333	27,917	1.2	263	27,002	1.0
- Other	126	17,525	0.7	79	9,161	0.9	47	8,364	0.6
NYS Excluding NYC									
- All Races	1,434	140,661	1.0	819	72,131	1.1	615	68,530	0.9
- White	1,204	121,069	1.0	691	62,125	1.1	513	58,944	0.9
- Black	187	14,544	1.3	98	7,354	1.3	89	7,190	1.2
- Other	36	4,273	0.8	25	2,244	1.1	11	2,029	0.5
New York City									
- All Races	1,093	122,950	0.9	626	62,953	1.0	467	59,997	0.8
- White	587	68,247	0.9	332	34,935	1.0	255	33,312	0.8
- Black	409	40,375	1.0	235	20,563	1.1	174	19,812	0.9
- Other	90	13,252	0.7	54	6,917	0.8	36	6,335	0.6

Note: Total includes unknowns within each category, thus row and column figures may not sum to totals.

Section I - Table 4b
1997 Births – New York State Residents
Percent of Live Births With Two Or More Major Malformations
Sex by Race and Residence

	Both Sexes			Males			Females		
Race and Residence	Infants	Total Births	%	Infants	Total Births	%	Infants	Total Births	%
New York State									
- All Races	2,353	256,976	0.9	1,370	131,353	1.0	978	125,619	0.8
- White	1,688	183,073	0.9	999	93,469	1.1	685	89,601	0.8
- Black	544	54,948	1.0	298	28,042	1.1	246	26,906	0.9
- Other	117	17,847	0.7	71	9,286	0.8	45	8,560	0.5
NYS Excluding NYC									
- All Races	1,336	138,074	1.0	803	70,594	1.1	528	67,476	0.8
- White	1.139	118,658	1.0	691	60,694	1.1	444	57,961	0.8
- Black	164	14,467	1.1	91	7,375	1.2	73	7,092	1.0
- Other	33	4,404	0.7	21	2,247	0.9	11	2,156	0.5
New York City									
- All Races	1,017	118,902	0.9	567	60,759	0.9	450	58,143	0.8
- White	549	64,415	0.9	308	32,775	0.9	241	31,640	0.8
- Black	380	40,481	0.9	207	20,667	1.0	173	19,814	0.9
- Other	84	13,443	0.6	50	7,039	0.7	34	6,404	0.5

Note: Total includes unknowns within each category, thus row and column figures may not sum to totals.

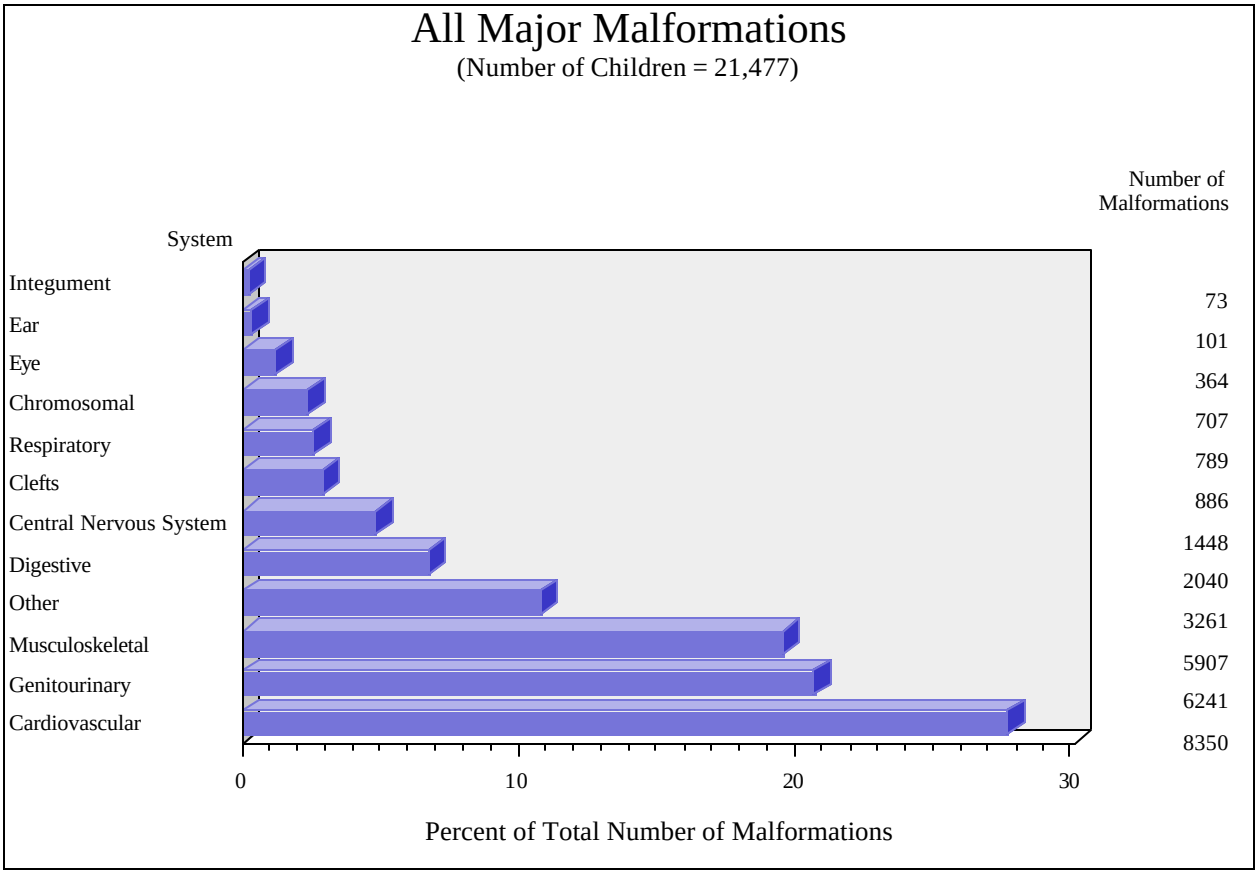
NOTES

Section II
Major Congenital Malformations by Organ System

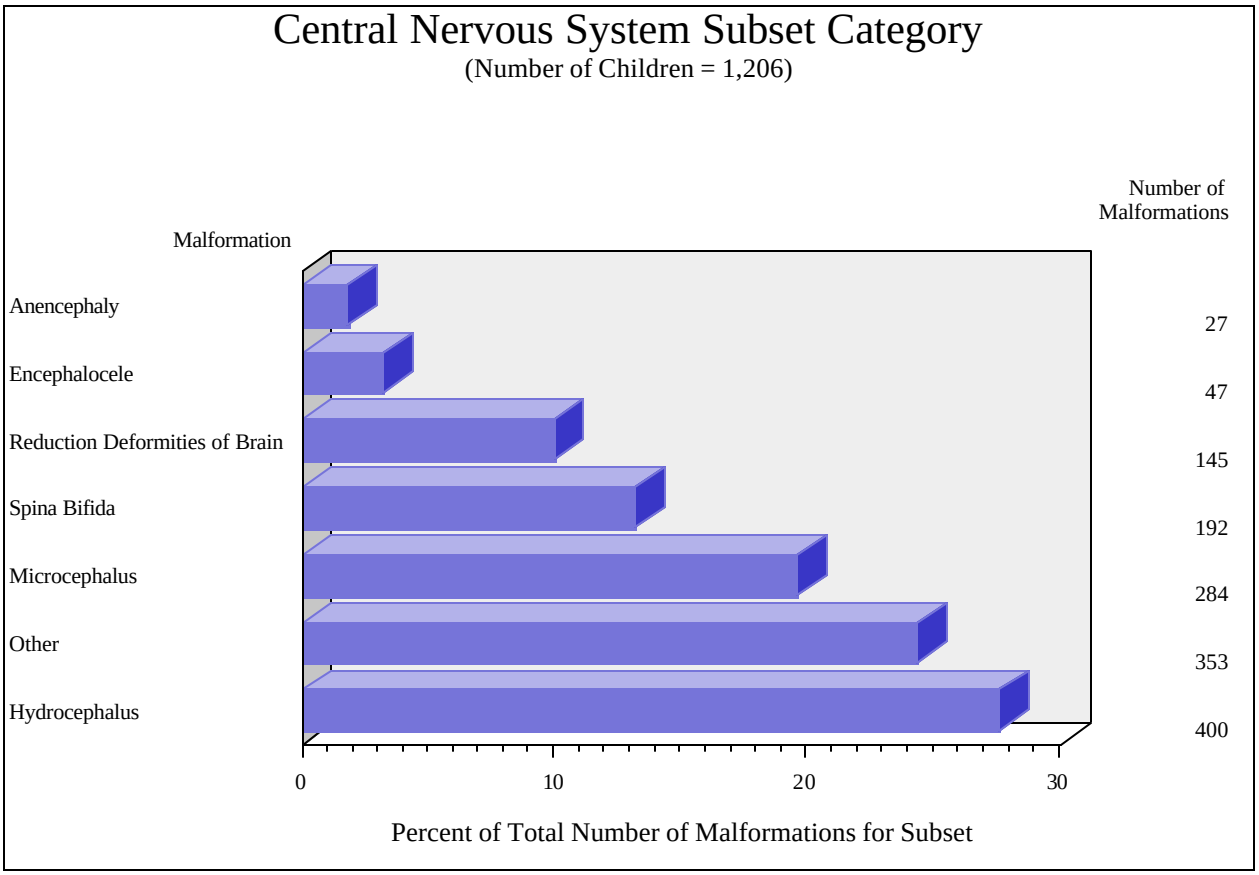
Introduction to Figures

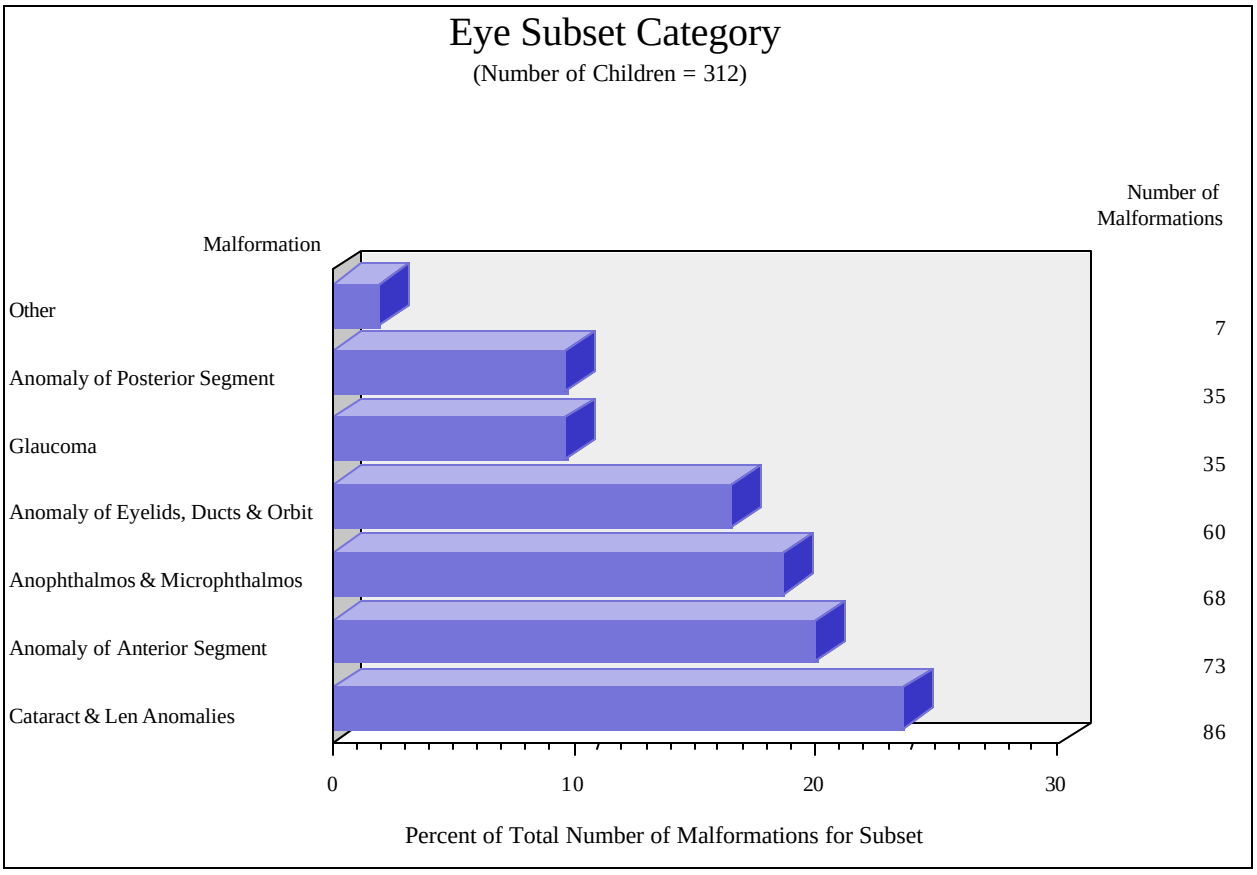
The organ system figures in this section present the distribution of 12 categories of major malformations, the relative contribution of each category to overall prevalence of major malformations in New York State, and the contribution of type of malformation within each subset category. Some of these percentages may differ from previous reports because of the new malformation coding system described in the Program Overview.

Major Malformation by Organ System
1996 & 1997 Births – New York State Residents

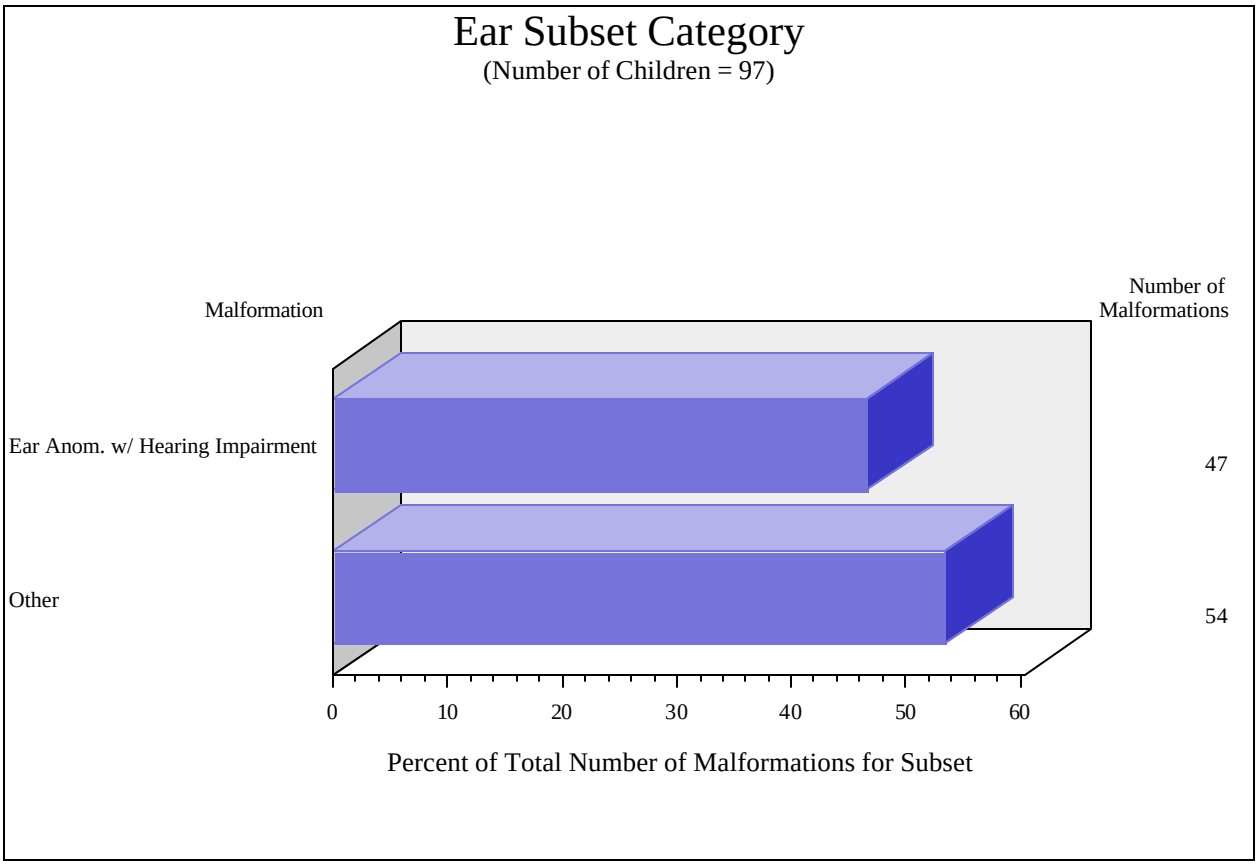


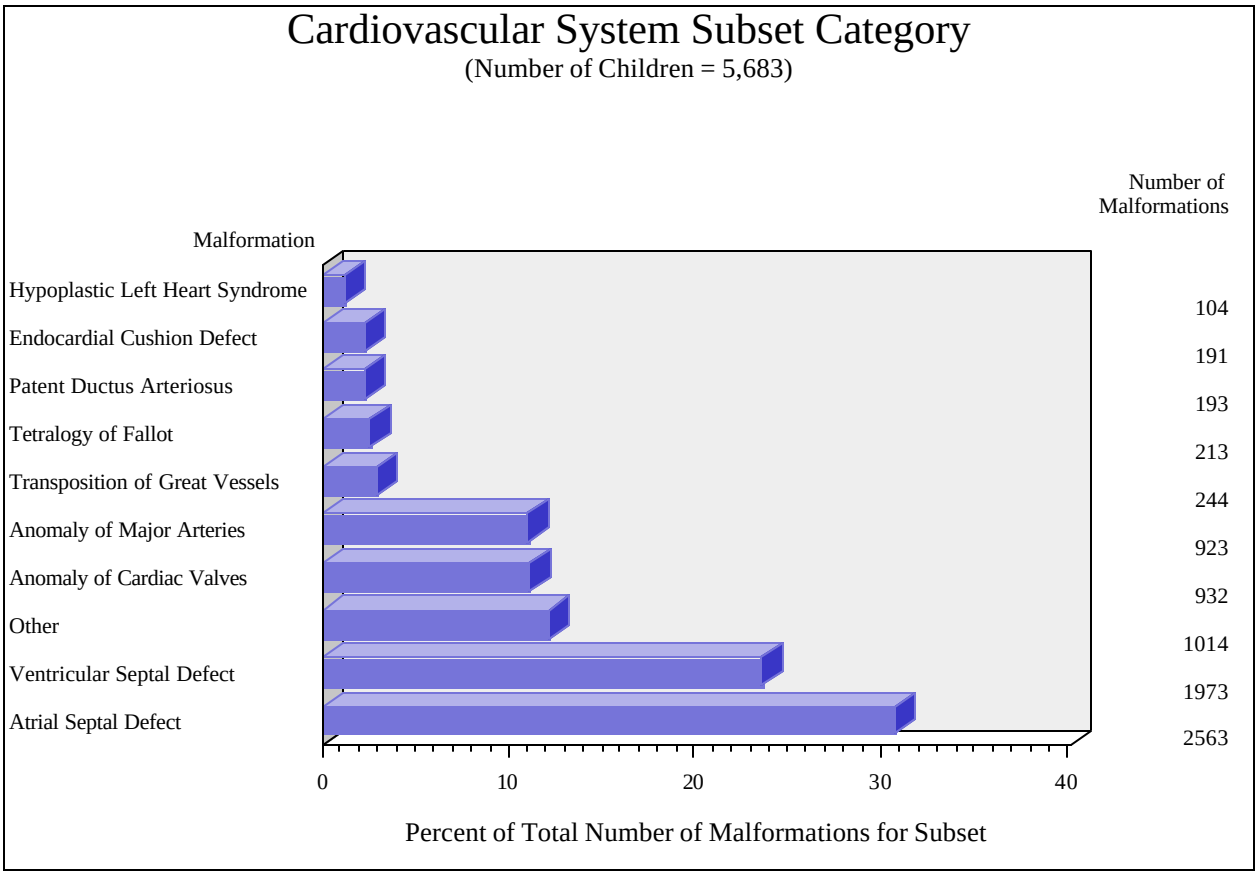
Major Malformations by Organ System
1996 & 1997 Births – New York State Residents



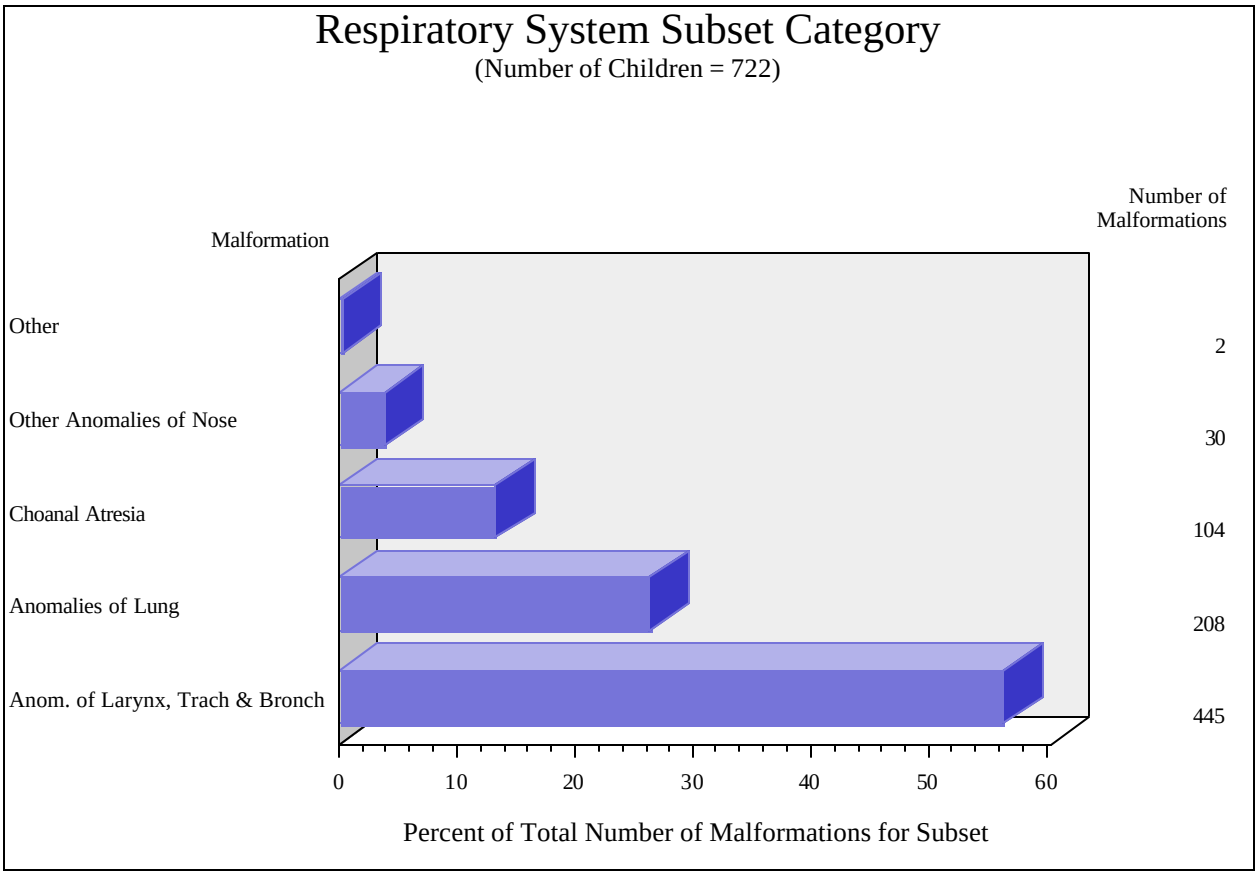


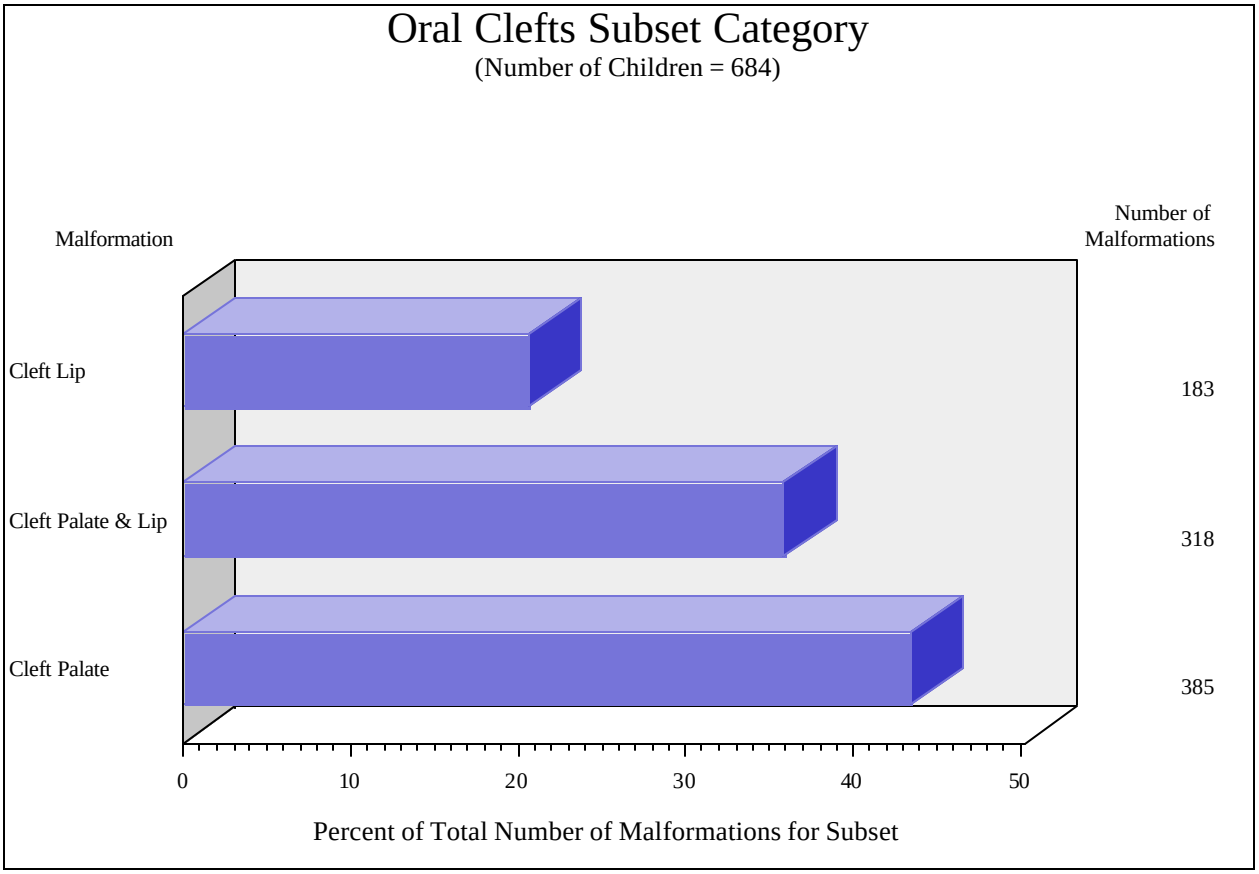
Major Malformations by Organ System
1996 & 1997 Births – New York State Residents



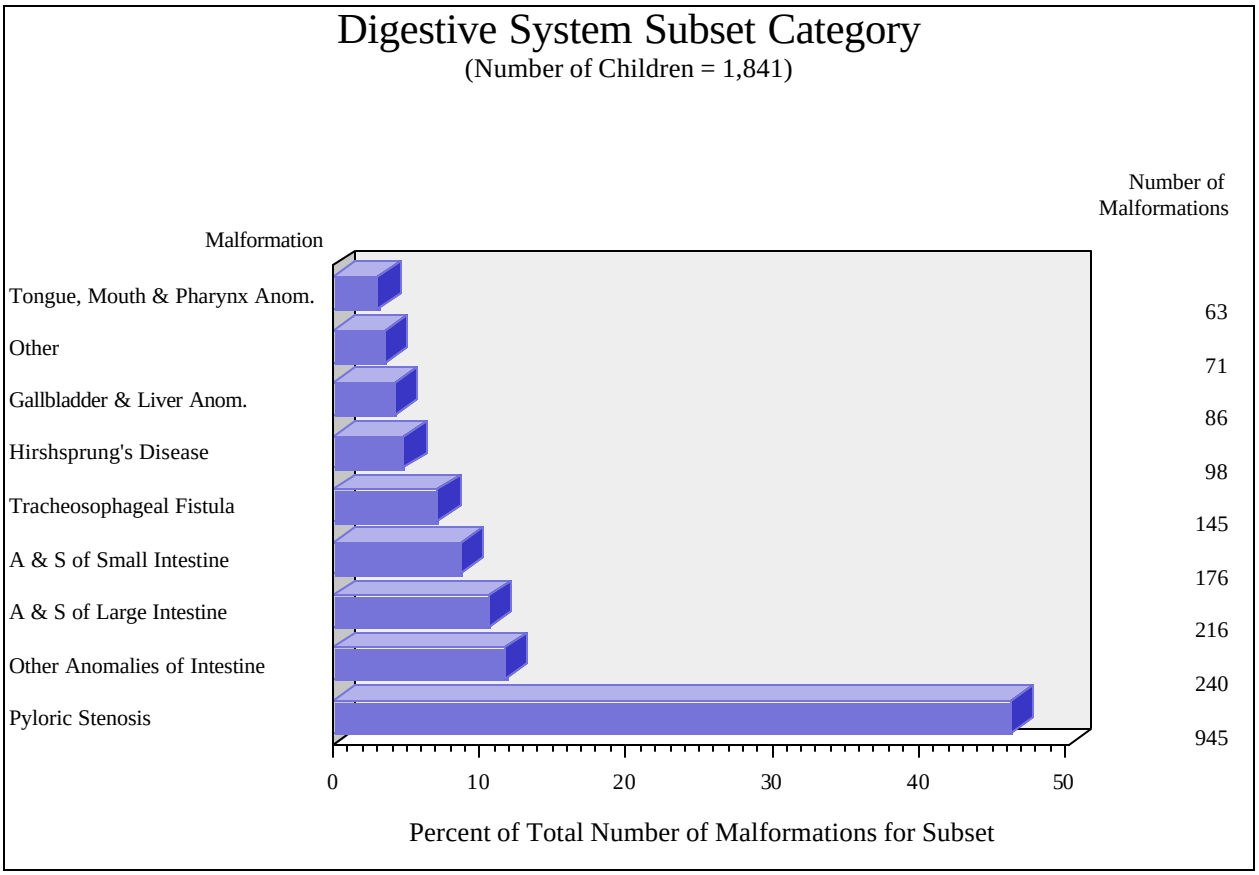


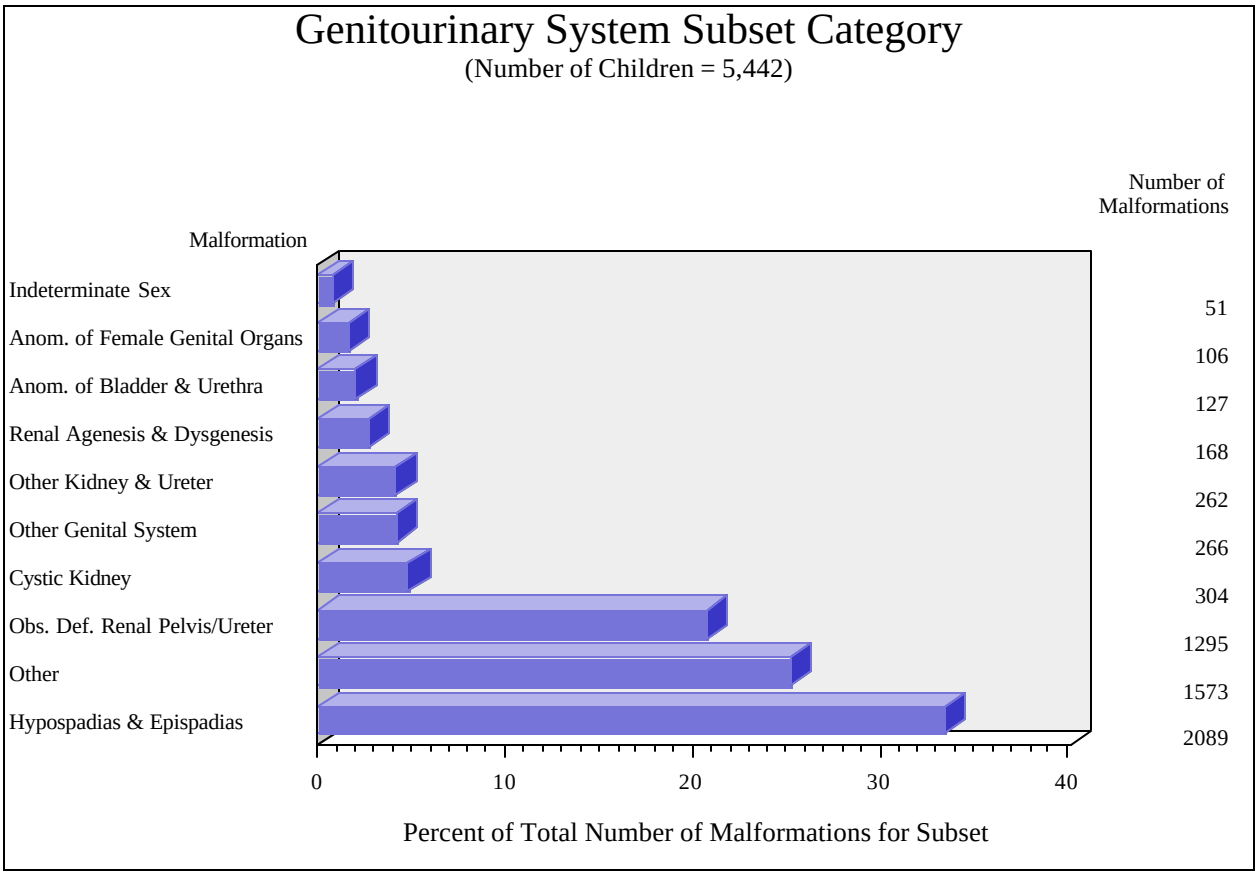
Major Malformations by Organ System
1996 & 1997 Births – New York State Residents



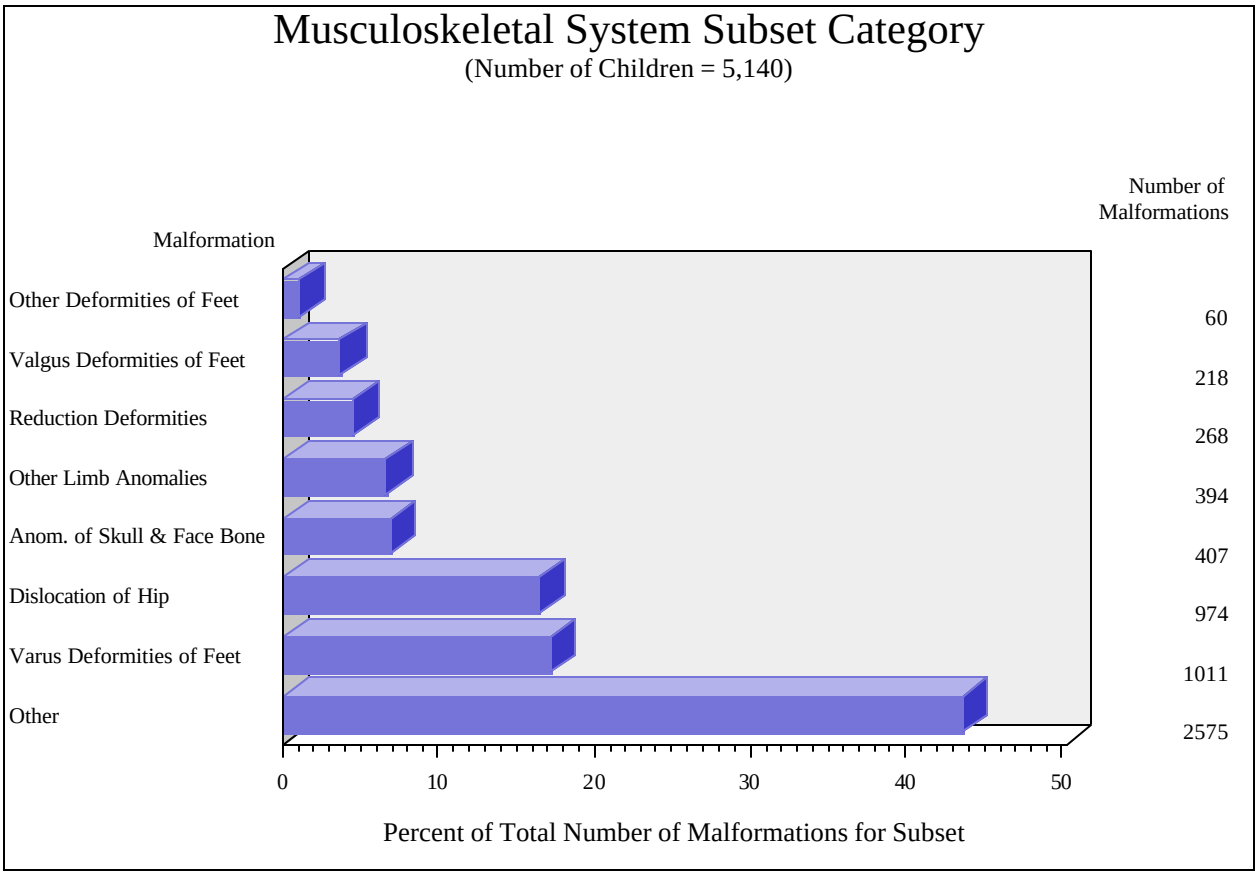


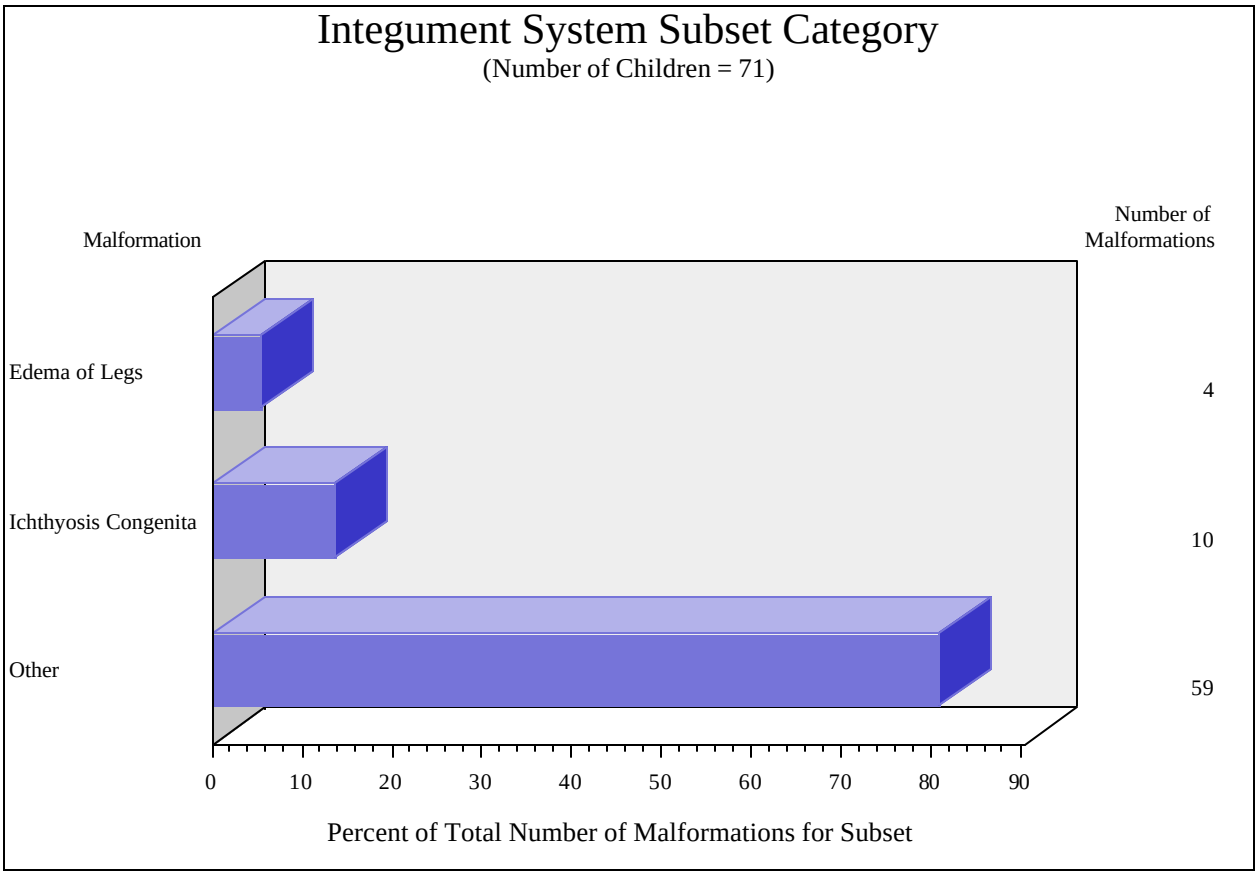
Major Malformations by Organ System
1996 & 1997 Births – New York State Residents



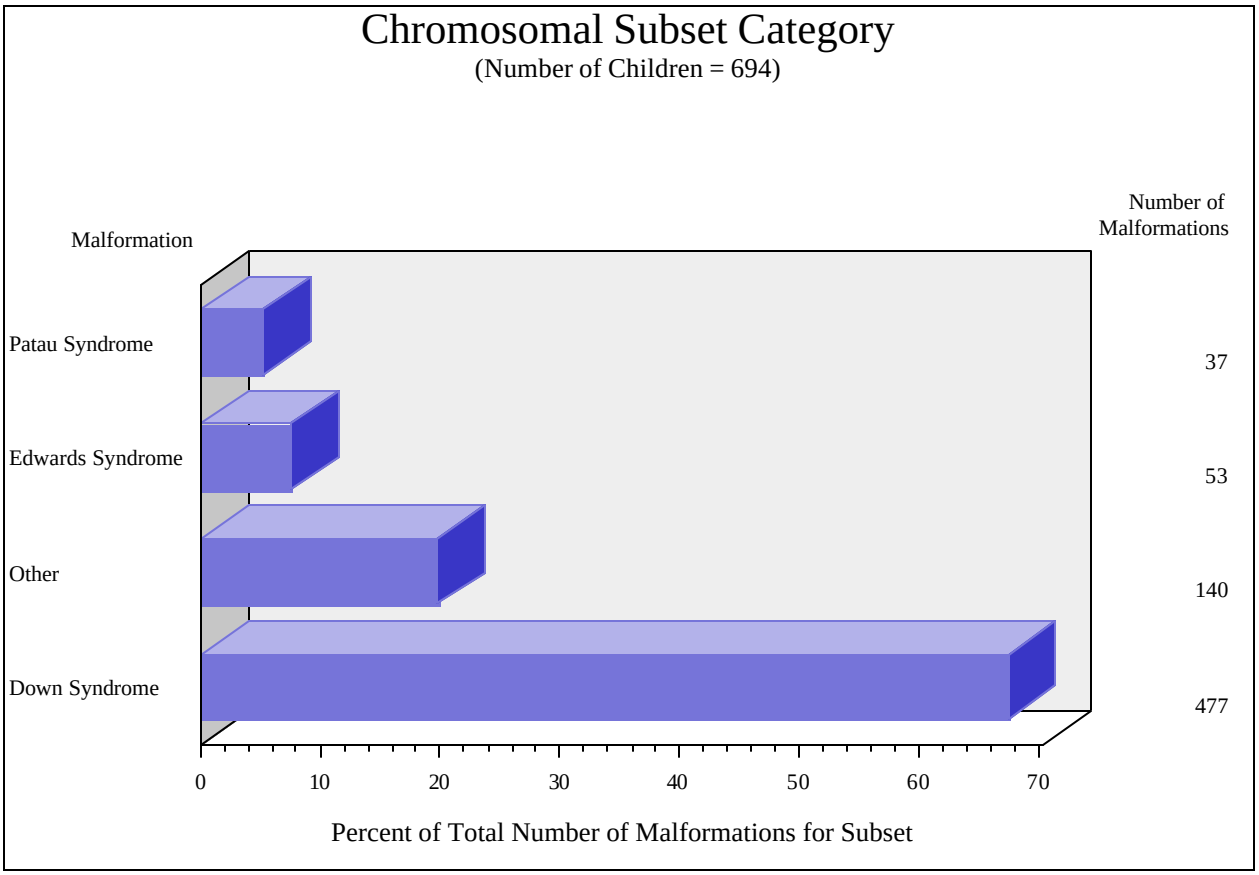


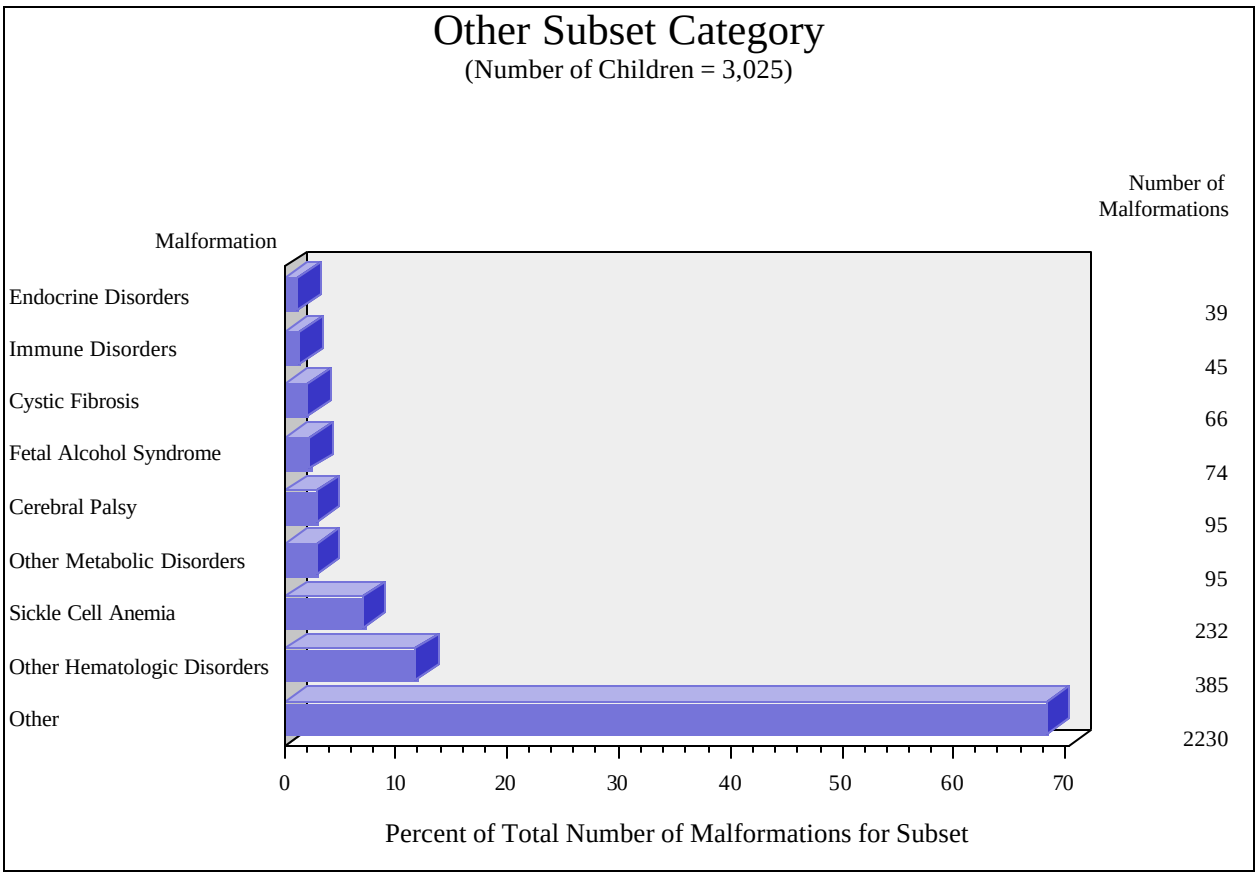
Major Malformations by Organ System
1996 & 1997 Births – New York State Residents





Major Malformations by Organ System
1996 & 1997 Births – New York State Residents





NOTES

Section III
Prevalence of Selected Malformations By Sex and Race

Introduction to Tables

The malformations presented in this section were selected because of the frequency with which they were reported and/or their clinical significance. Rates are per 10,000 live births. The sex ratio is calculated by dividing the rate in males by the rate in females. The malformation rates presented in this report may not be comparable to earlier reports. Previous reports from 1989 to 1991 did not use birth certificate matched cases; thus, the race and birthweight from the birth certificate were not available. Birthweight data are useful to calculate the rate of some malformations such as patent ductus arteriosus. In some cases, these conditions can result from being preterm rather than actually having a malformation. Racial data in this report also may not be comparable because race is defined by maternal race from the birth certificate. In the earlier reports, race was defined by what was reported on the CMR form, which may differ from what is recorded on the birth certificate.

Fluctuations in specific malformation prevalence should be interpreted with caution, especially differences in the "other" race category since the numbers in this group are small. In addition, several malformations were added in 1992 as a result of the change to the BPA code. Previously, these could not be distinguished using the ICD-9 codes. However, since ICD-9 codes are more familiar to most vendors, the ICD-9 code is given on the table with the named malformation. See Appendix 4 for further information on the BPA codes.

Section III
Children with Selected Major Malformations
Prevalences per 10,000 Live Births by Sex & Race

1996 Births – New York State Residents

ICD-9 Code	Malformation	Total Number	Total Prevalence	Male	Female	Ratio (M/F)	White	Black	Other
243	Congenital hypothyroidism	72	2.7	2.7	2.8	1.0	2.2	4.6	2.9
270.1	Phenylketonuria	2	0.1	0.1	0.1	1.0	0.1	0.0	0.0
277.0	Cystic fibrosis	30	1.1	1.4	0.9	1.6	1.5	0.2	0.6
282.6	Sickle-cell anemia	123	4.7	4.1	5.3	0.8	0.4	20.8	0.0
740.0	Anencephalus	16	0.6	0.4	0.8	0.6	0.6	0.5	0.6
741.0	Spina bifida with hydrocephalus	47	1.8	1.8	1.8	1.0	2.2	0.7	1.1
741.9	Spina bifida without hydrocephalus	45	1.7	1.8	1.6	1.1	1.8	1.3	2.3
742.0	Encephalocele	25	0.9	0.8	1.1	0.7	1.1	0.5	0.6
742.1	Microcephalus	152	5.8	4.7	6.8	0.7	4.9	9.1	5.7
742.2	Agyria & lissencephaly	10	0.4	0.3	0.5	0.6	0.2	1.1	0.0
742.2	Anomalies of corpus callosum	46	1.7	1.9	1.6	1.2	2.0	1.3	0.6
742.2	Holoprosencephaly	9	0.3	0.1	0.5	0.3	0.4	0.2	0.6
742.3	Congenital hydrocephalus	206	7.8	7.6	8.1	0.9	7.4	10.6	2.9
742.4	Porencephaly	19	0.7	0.7	0.8	0.9	0.6	1.3	0.6
742.59	Congenital tethered cord	22	0.8	1.0	0.7	1.4	1.0	0.2	1.1
743.0	Anophthalmos	3	0.1	0.1	0.1	1.9	0.1	0.2	0.0
743.1	Microphthalmos	34	1.3	1.3	1.2	1.1	1.4	1.3	0.6
743.2	Glaucoma	14	0.5	0.6	0.5	1.3	0.5	0.7	0.6
743.5	Absence of lens	3	0.1	0.1	0.2	0.5	0.2	0.0	0.0
743.3	Congenital cataract	35	1.3	1.3	1.4	0.9	1.3	1.6	1.1
743.45	Aniridia	5	0.2	0.1	0.3	0.2	0.2	0.4	0.0
743.46	Coloboma of iris	10	0.4	0.4	0.4	1.0	0.5	0.2	0.0
744.0,2	Anotia/microtia	13	0.5	0.6	0.4	1.5	0.7	0.0	0.0
745.0	Common truncus	18	0.7	0.5	0.9	0.6	0.7	0.9	0.0
745.1	Transposition of great vessels	105	4.0	4.7	3.3	1.4	4.1	4.2	1.1
745.2	Tetralogy of Fallot	112	4.2	4.7	3.7	1.3	4.3	4.6	2.9
745.3	Common ventricle	23	0.9	0.7	1.0	0.7	1.0	0.9	0.0
745.4	Ventricular septal defect	1023	38.8	35.1	42.7	0.8	39.6	36.1	37.1
745.5	Ostium secundum type atrial septal def.	1199	45.5	44.0	47.0	0.9	40.0	67.2	37.1
745.6	Endocardial cushion defects	74	2.8	3.2	2.4	1.3	2.7	2.9	3.4
746.0	Atresia/stenosis of pulmonary valve	274	10.4	10.7	10.1	1.1	9.0	16.0	6.8
746.1	Tricuspid atresia/stenosis/hypoplasia	28	1.1	1.0	1.1	1.0	0.8	2.0	0.6
746.2	Ebstein's anomaly	9	0.3	0.3	0.4	0.8	0.3	0.2	0.6
746.3	Congenital stenosis of aortic valve	37	1.4	1.9	0.9	2.2	1.8	0.2	0.6
746.7	Hypoplastic left heart syndrome	53	2.0	1.8	2.3	0.8	1.8	3.3	0.0
746.85	Anomalies of coronary artery	12	0.5	0.3	0.6	0.5	0.5	0.2	0.0
747.0	Patent ductus arteriosus	95	3.6	3.4	3.8	0.9	2.8	5.8	5.1
747.10	Coarctation of aorta	114	4.3	4.7	3.9	1.2	4.6	3.8	3.4
747.41	Total anomalous pulmonary venous connect.	32	1.2	1.3	1.1	1.2	1.0	1.8	1.7
748.0	Choanal atresia	59	2.2	2.0	2.5	0.8	2.8	0.9	0.6
748.5	Agenesis/hypoplasia of lung	73	2.8	2.7	2.8	1.0	3.0	2.5	1.1
749.0	Cleft palate	161	6.1	4.9	7.4	0.7	6.3	5.1	8.0
749.1	Cleft lip	87	3.3	3.9	2.6	1.5	3.8	1.6	3.4
749.2	Cleft palate & lip	129	4.9	6.4	3.3	2.0	5.3	3.1	5.7
750.3	Tracheoesophageal fistula etc.	64	2.4	2.8	2.0	1.4	2.5	2.4	1.7
750.5	Congenital hypertrophic pyloric stenosis	469	17.8	28.4	6.7	4.2	21.4	8.6	7.4

Section III
Children with Selected Major Malformations
Prevalences per 10,000 Live Births by Sex & Race

1996 Births – New York State Residents (continued)

ICD-9 Code	Malformation	Total Number	Total Prevalence	Male	Female	Ratio (M/F)	White	Black	Other
751.1	Atresia and stenosis of small intestine	80	3.0	2.7	3.3	0.8	2.7	4.4	2.3
751.2	Atresia and stenosis of rectum or anus	88	3.3	3.7	3.0	1.3	3.7	2.2	2.9
751.3	Hirschsprungs disease	59	2.2	3.0	1.5	2.0	2.1	3.3	1.1
751.4	Anomalies of intestinal fixation	52	2.0	2.2	1.7	1.3	2.1	2.0	0.6
751.61	Biliary atresia	26	1.0	1.3	0.7	1.8	0.8	0.7	3.4
752.6	Epispadias	16	0.6	1.2	0.0		0.6	0.9	0.0
752.6	Hypospadias	883	33.5	64.6	0.9	75.4	37.2	24.6	22.3
753.0	Renal agenesis and dysgenesis	79	3.0	3.4	2.6	1.3	3.5	2.0	0.6
753.1	Cystic kidney disease	143	5.4	5.3	5.6	0.9	5.9	4.9	2.9
753.2	Obstructive defect renal pelvis & ureter	624	23.7	31.2	15.7	2.0	25.0	17.8	28.0
753.5	Exstrophy of urinary bladder	9	0.3	0.4	0.2	1.9	0.3	0.4	0.0
753.6	Atresia & stenosis of urethra & bladder	43	1.6	3.1	0.1	40.0	1.2	2.5	3.4
754.3	Congenital dislocation of hip	347	13.2	6.7	20.0	0.3	14.5	8.0	14.3
754.51	Talipes equinovarus	329	12.5	15.8	8.9	1.8	14.1	8.7	7.4
755.2	Reduction deformities of upper limb	71	2.7	3.0	2.3	1.3	3.1	1.8	1.1
755.3	Reduction deformities of lower limb	36	1.4	1.3	1.5	0.9	1.1	2.5	0.6
754.89	Arthrogryposis multiplex congenita	31	1.2	1.1	1.2	0.9	1.3	0.9	0.0
756.0	Craniosynostosis	102	3.9	4.1	3.6	1.2	4.6	1.8	2.9
756.0	Goldenhar syndrome	11	0.4	0.7	0.2	4.3	0.5	0.2	0.0
756.4	Chondrodystrophy	24	0.9	1.0	0.8	1.3	0.9	1.1	0.6
756.51	Osteogenesis imperfecta	9	0.3	0.3	0.4	0.8	0.4	0.2	0.0
756.6	Diaphragmatic hernia	62	2.4	2.7	2.0	1.3	2.2	2.5	2.3
756.7	Gastroschisis	43	1.6	1.6	1.7	0.9	1.8	1.3	0.6
756.7	Omphalocele	37	1.4	1.3	1.5	0.9	1.1	2.7	0.6
756.7	Prune belly	4	0.2	0.2	0.1	2.9	0.1	0.5	0.0
758.0	Down syndrome	234	8.9	8.6	9.2	0.9	9.0	8.9	8.0
758.1	Patau syndrome	13	0.5	0.6	0.4	1.5	0.4	0.7	1.1
758.2	Edwards syndrome	22	0.8	0.7	0.9	0.8	0.8	1.1	0.0
758.6	Gonadal dysgenesis	16	0.6	0.1	1.1	0.1	0.8	0.0	0.0
758.7	Klinefelter syndrome	14	0.5	1.0	0.0		0.5	0.5	0.6
759.3	Situs inversus	17	0.6	0.9	0.4	2.3	0.6	0.9	0.0
760.71	Fetal alcohol syndrome	41	1.6	2.0	1.1	1.8	0.7	4.9	0.0
771.0	Congenital rubella	4	0.2	0.0	0.3	0.0	0.1	0.4	0.0
771.1	Congenital cytomegalovirus infection	28	1.1	1.0	1.1	1.0	0.5	3.3	0.6
771.2	Other congenital infections	40	1.5	1.8	1.2	1.4	1.5	2.2	0.0

Section III
Children with Selected Major Malformations
Prevalences per 10,000 Live Births by Sex & Race

1997 Births – New York State Residents

ICD-9 Code	Malformation	Total Number	Total Prevalence	Male	Female	Ratio (M/F)	White	Black	Other
243	Congenital hypothyroidism	65	2.5	3.2	1.8	1.7	2.1	3.5	3.9
270.1	Phenylketonuria	2	0.1	0.0	0.2	0.0	0.0	0.4	0.0
277.0	Cystic fibrosis	34	1.3	1.3	1.4	1.0	1.4	1.5	0.0
282.6	Sickle-cell anemia	98	3.8	3.8	3.8	1.0	0.8	14.9	0.6
740.0	Anencephalus	11	0.4	0.4	0.5	0.8	0.4	0.4	0.6
741.0	Spina bifida with hydrocephalus	36	1.4	1.4	1.4	1.0	1.3	1.6	2.2
741.9	Spina bifida without hydrocephalus	37	1.4	1.3	1.6	0.8	1.6	0.7	1.7
742.0	Encephalocele	20	0.8	0.9	0.6	1.4	0.6	1.5	0.6
742.1	Microcephalus	132	5.1	4.0	6.3	0.6	3.8	8.9	7.3
742.2	Agyria & lissencephaly	8	0.3	0.3	0.3	1.0	0.3	0.2	1.1
742.2	Anomalies of corpus callosum	41	1.6	1.6	1.6	1.0	1.5	1.5	2.8
742.2	Holoprosencephaly	10	0.4	0.5	0.3	1.4	0.5	0.2	0.0
742.3	Congenital hydrocephalus	184	7.2	8.1	6.2	1.3	6.9	7.8	8.4
742.4	Porencephaly	11	0.4	0.6	0.2	2.6	0.5	0.2	0.6
742.59	Congenital tethered cord	21	0.8	0.5	1.1	0.5	1.0	0.5	0.0
743.0	Anophthalmos	3	0.1	0.1	0.2	0.5	0.1	0.4	0.0
743.1	Microphthalmos	27	1.1	0.8	1.3	0.7	1.0	1.3	1.1
743.2	Glaucoma	19	0.7	1.1	0.3	3.6	0.7	0.9	1.1
743.3	Congenital cataract	36	1.4	1.5	1.2	1.3	1.5	0.9	2.2
743.45	Aniridia	5	0.2	0.2	0.2	1.4	0.2	0.4	0.0
743.46	Coloboma of iris	9	0.4	0.4	0.3	1.2	0.3	0.4	0.6
744.0,2	Anotia/microtia	10	0.4	0.4	0.4	1.0	0.5	0.0	0.6
745.0	Common truncus	20	0.8	0.8	0.8	1.0	0.8	0.9	0.6
745.1	Transposition of great vessels	96	3.7	4.8	2.6	1.8	4.0	3.3	2.2
745.2	Tetralogy of Fallot	100	3.9	3.8	3.9	1.0	4.1	3.5	3.4
745.3	Common ventricle	20	0.8	0.8	0.8	1.0	1.0	0.4	0.0
745.4	Ventricular septal defect	950	37.0	35.4	38.4	0.9	38.3	34.6	31.9
745.5	Ostium secundum type atrial septal def.	1241	48.3	47.8	48.7	1.0	44.5	65.2	35.9
745.6	Endocardial cushion defects	87	3.4	3.0	3.7	0.8	3.2	4.5	2.2
746.0	Atresia/stenosis of pulmonary valve	222	8.6	8.1	9.2	0.9	8.0	12.2	5.0
746.1	Tricuspid atresia/stenosis/hypoplasia	35	1.4	1.1	1.6	0.7	1.4	1.6	0.0
746.2	Ebstein's anomaly	16	0.6	0.5	0.7	0.7	0.8	0.4	0.0
746.3	Congenital stenosis of aortic valve	33	1.3	1.6	1.0	1.7	1.5	0.7	0.6
746.7	Hypoplastic left heart syndrome	51	2.0	2.7	1.3	2.1	1.9	2.9	0.0
746.85	Anomalies of coronary artery	7	0.3	0.4	0.2	2.4	0.3	0.4	0.0
747.0	Patent ductus arteriosus	98	3.8	3.8	3.8	1.0	3.4	4.7	5.0
747.10	Coarctation of aorta	100	3.9	5.2	2.5	2.1	4.4	3.3	1.1
747.41	Total anomalous pulmonary venous connect.	21	0.8	0.9	0.7	1.3	0.9	0.7	0.0
748.0	Choanal atresia	45	1.8	1.8	1.8	1.0	2.1	1.3	0.0
748.5	Agenesis/hypoplasia of lung	89	3.5	3.8	3.1	1.2	3.3	4.4	2.2
749.0	Cleft palate	166	6.5	5.7	7.2	0.8	7.0	3.8	9.0
749.1	Cleft lip	69	2.7	2.7	2.7	1.0	2.9	1.5	3.9
749.2	Cleft palate & lip	124	4.8	5.5	4.1	1.3	5.2	2.7	6.2
750.3	Tracheoesophageal fistula etc.	57	2.2	2.1	2.4	0.9	2.3	2.2	1.7
750.5	Congenital hypertrophic pyloric stenosis	472	18.4	28.5	7.8	3.6	22.2	9.3	6.7
751.1	Atresia and stenosis of small intestine	83	3.2	2.5	4.0	0.6	3.2	3.8	1.7
751.2	Atresia and stenosis of rectum or anus	98	3.8	3.9	3.7	1.0	4.3	2.0	4.5
751.3	Hirschsprungs disease	39	1.5	2.1	1.0	2.2	1.6	1.5	0.6
751.4	Anomalies of intestinal fixation	41	1.6	1.8	1.4	1.4	2.0	0.9	0.0
751.61	Biliary atresia	21	0.8	0.7	1.0	0.7	0.8	0.9	0.6
752.6	Epispadias	13	0.5	1.0	0.0		0.6	0.4	0.0
752.6	Hypospadias	817	31.8	62.0	0.1	778.5	35.1	24.2	21.3
753.0	Renal agenesis and dysgenesis	83	3.2	3.7	2.7	1.4	3.8	2.2	1.1
753.1	Cystic kidney disease	130	5.1	5.8	4.3	1.3	4.6	7.5	2.8

Section III
Children with Selected Major Malformations
Prevalences per 10,000 Live Births by Sex & Race

1997 Births – New York State Residents (continued)

ICD-9 Code	Malformation	Total Number	Total Prevalence	Male	Female	Ratio (M/F)	White	Black	Other
753.2	Obstructive defect renal pelvis & ureter	546	21.2	31.2	10.8	2.9	22.6	16.0	24.1
753.5	Exstrophy of urinary bladder	2	0.1	0.1	0.1	1.0	0.1	0.0	0.0
753.6	Atresia & stenosis of urethra & bladder	31	1.2	2.3	0.1	28.7	1.1	1.5	1.1
754.3	Congenital dislocation of hip	340	13.2	6.5	20.2	0.3	15.2	6.2	15.1
754.51	Talipes equinovarus	297	11.6	15.2	7.8	1.9	12.3	11.1	6.2
755.2	Reduction deformities of upper limb	72	2.8	2.2	3.4	0.6	3.2	2.4	0.6
755.3	Reduction deformities of lower limb	38	1.5	1.5	1.4	1.1	1.4	2.0	1.1
754.89	Arthrogryposis multiplex congenita	35	1.4	1.4	1.3	1.1	1.3	1.5	1.7
756.0	Craniosynostosis	88	3.4	4.0	2.8	1.4	4.1	1.6	2.2
756.0	Goldenhar syndrome	8	0.3	0.3	0.3	1.0	0.4	0.0	0.0
756.4	Chondrodystrophy	21	0.8	1.0	0.6	1.6	0.9	0.9	0.0
756.51	Osteogenesis imperfecta	7	0.3	0.4	0.2	2.4	0.2	0.5	0.0
756.6	Diaphragmatic hernia	40	1.6	2.0	1.0	1.9	1.7	1.5	0.6
756.7	Gastroschisis	35	1.4	1.5	1.2	1.3	1.4	1.3	1.1
756.7	Omphalocele	36	1.4	1.4	1.4	1.0	1.1	2.0	1.7
756.7	Prune belly	3	0.1	0.2	0.0		0.1	0.4	0.0
758.0	Down syndrome	239	9.3	9.3	9.3	1.0	9.9	8.0	7.3
758.1	Patau syndrome	24	0.9	1.0	0.9	1.1	0.9	1.1	0.6
758.2	Edwards syndrome	31	1.2	1.1	1.3	0.9	1.4	0.9	0.0
758.6	Gonadal dysgenesis	10	0.4	0.1	0.7	0.1	0.5	0.0	0.0
758.7	Klinefelter syndrome	5	0.2	0.4	0.0		0.2	0.2	0.0
759.3	Situs inversus	17	0.7	0.6	0.7	0.9	0.6	0.9	0.6
760.71	Fetal alcohol syndrome	33	1.3	1.4	1.1	1.3	0.4	4.4	0.0
771.0	Congenital rubella	4	0.2	0.2	0.2	1.0	0.1	0.2	1.1
771.1	Congenital cytomegalovirus infection	27	1.1	1.1	1.0	1.0	0.8	2.0	0.6
771.2	Other congenital infections	41	1.6	2.1	1.0	2.1	1.1	2.4	3.9

NOTES

Section IV
Most Frequently Reported Selected Major Malformations by County

Introduction to Tables

Congenital Malformation Registry data were tabulated by county of residence at the time of birth and four digit ICD-9-CM codes for major malformations. Certain codes for rare disorders and nonspecific codes are not included. The table on the next page presents the number of children with major malformations by county, and the total number of live births for comparison.

The maps of New York State showing county live birth rates (Figures 4a, 4b) are part of the Bureau of Biometrics annual reports, "Vital Statistics of New York State 1996 & 1997." The Bureau of Biometrics supplies the CMR with the number of live births in a specific year, which is used as a denominator in the calculation of prevalences of malformations.

For each county, the 10 most frequently reported codes are listed, except those instances in which the tenth and subsequent codes were equal in number. In this circumstance, the additional codes of equal number are listed. Some counties may have fewer than 10 codes reported. Children reported with more than one malformation may be represented more than once in these tables. These are presented on the following pages.

These county listings are not designed to be used for comparison among counties or for analytic studies. They are most useful to assist in county planning, education, counseling and other health care services programs.

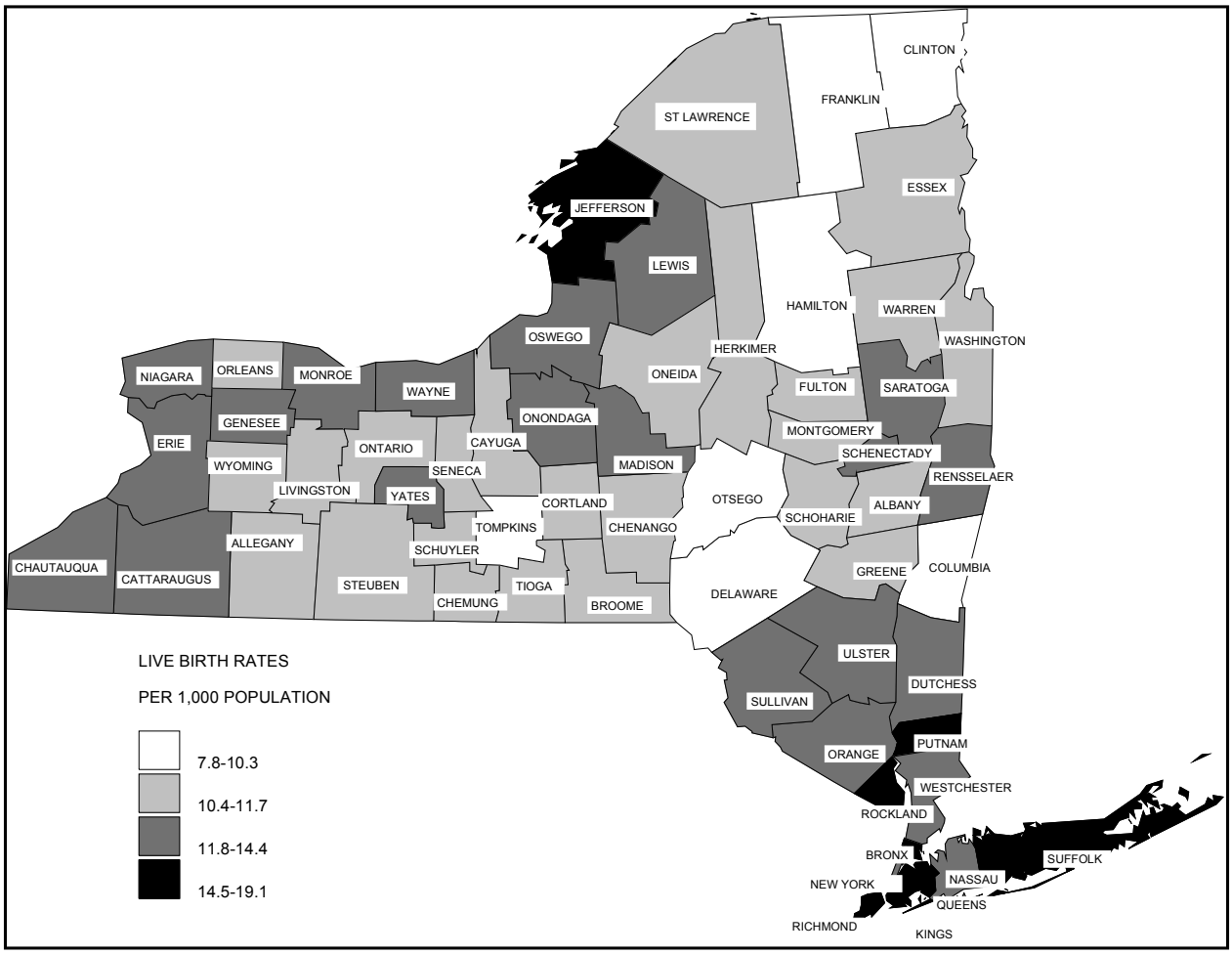
For information about vital statistics, contact:

New York State Department of Health
Bureau of Biometrics
ESP Concourse - C144
Albany, NY 12237-0044.

Children with Major Congenital Malformations and Births by County, 1996-1997

	1996			1997		
County	Number of Children	Number of Live Births	Precent of Live Births	Number of Children	Number of Live Births	Percent of Live Births
Albany	137	3,307	4.1	138	3,276	4.2
Allegany	20	581	3.4	30	560	5.4
Bronx	1,017	23,404	4.3	867	22,457	3.9
Broome	113	2,258	5.0	105	2,201	4.8
Cattaraugus	61	1,109	5.5	42	1,046	4.0
Cayuga	40	991	4.0	29	933	3.1
Chautauqua	69	1,689	4.1	73	1,624	4.5
Chemung	53	1,078	4.9	56	1,037	5.4
Chenango	20	621	3.2	18	609	3.0
Clinton	32	891	3.6	33	795	4.2
Columbia	28	670	4.2	22	664	3.3
Cortland	18	583	3.1	20	562	3.6
Delaware	14	454	3.1	23	491	4.7
Dutchess	117	3,348	3.5	138	3,399	4.1
Erie	699	12,031	5.8	718	11,635	6.2
Essex	7	413	1.7	7	391	1.8
Franklin	13	510	2.5	13	465	2.8
Fulton	38	641	5.9	30	626	4.8
Genesee	33	753	4.4	40	755	5.3
Greene	13	499	2.6	18	491	3.7
Hamilton	2	43	4.7	0	46	0.0
Herkimer	31	704	4.4	38	700	5.4
Jefferson	60	1,793	3.3	54	1,734	3.1
Kings	1,810	40,928	4.4	1,695	39,746	4.3
Lewis	25	369	6.8	18	336	5.4
Livingston	19	712	2.7	27	706	3.8
Madison	39	858	4.5	35	826	4.2
Monroe	352	9,669	3.6	330	9,622	3.4
Montgomery	29	575	5.0	27	594	4.5
Nassau	823	17,722	4.6	858	17,100	5.0
New York	687	20,045	3.4	642	19,366	3.3
Niagara	139	2,744	5.1	133	2,641	5.0
Oneida	123	2,702	4.6	134	2,702	5.0
Onondaga	315	6,283	5.0	232	5,972	3.9
Ontario	55	1,146	4.8	60	1,180	5.1
Orange	160	4,893	3.3	159	4,869	3.3
Orleans	30	526	5.7	24	550	4.4
Oswego	79	1,509	5.2	57	1,445	3.9
Otsego	18	549	3.3	17	586	2.9
Putnam	33	1,282	2.6	42	1,227	3.4
Queens	1,248	32,691	3.8	1,227	31,623	3.9
Rensselaer	67	1,945	3.4	65	1,784	3.6
Richmond	258	5,882	4.4	277	5,710	4.9
Rockland	139	4,239	3.3	126	4,341	2.9
Saratoga	97	2,523	3.8	97	2,405	4.0
Schenectady	76	1,777	4.3	58	1,750	3.3
Schoharie	12	364	3.3	15	341	4.4
Schuyler	12	206	5.8	5	205	2.4
Seneca	21	395	5.3	16	374	4.3
St. Lawrence	57	1,242	4.6	32	1,181	2.7
Steuben	56	1,136	4.9	53	1,186	4.5
Suffolk	836	19,953	4.2	788	19,862	4.0
Sullivan	38	839	4.5	22	839	2.6
Tioga	21	630	3.3	21	642	3.3
Tompkins	23	851	2.7	36	857	4.2
Ulster	59	1,976	3.0	61	1,922	3.2
Warren	29	673	4.3	19	689	2.8
Washington	33	695	4.7	22	610	3.6
Wayne	48	1,217	3.9	39	1,261	3.1
Westchester	483	12,696	3.8	443	12,655	3.5
Wyoming	26	471	5.5	24	443	5.4
Yates	10	327	3.1	9	331	2.7

Figure 4a: Live Birth Rates, New York State 1996



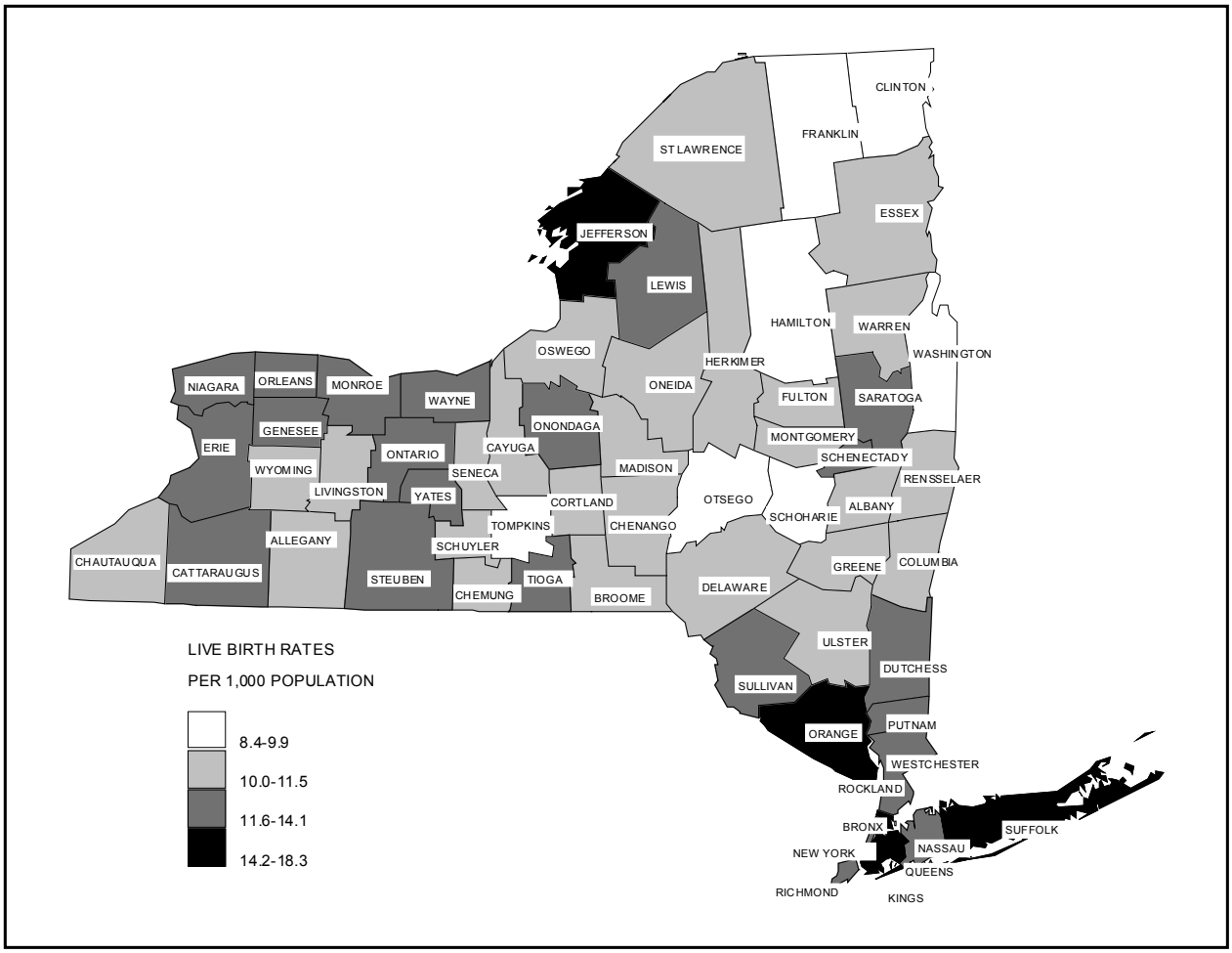
Highlights

- New York State has an overall live birth rate of 14.2 live births per 1,000 population in 1996, a decrease from 14.7 in 1995.

- The county with the highest live birth rate is Bronx County, with 19.1 live births per 1,000 population. Other counties with high live birth rates are Jefferson, Kings, Putnam, Queens, Richmond, Rockland, and Suffolk.

- Hamilton County has the lowest live birth rate in 1996, with 7.8 live births per 1,000 population. Low live birth rates are reported for Clinton, Columbia, Delaware, Franklin, Otsego, and Tompkins Counties.

Figure 4b: Live Birth Rates, New York State 1997



Highlights

- New York State has an overall live birth rate of 13.8 live births per 1,000 population in 1997, a decrease from 14.2 in 1996.

- The county with the highest live birth rate is Bronx County, with 18.3 live births per 1,000 population. Other counties with high live birth rates are Jefferson, Kings, Queens, Orange, Rockland, and Suffolk.

- Hamilton and Tompkins Counties have the lowest live birth rate in 1997, with 8.4 live births per 1,000 population. Low live birth rates are reported for Clinton, Franklin, Otsego, Schoharie, and Washington Counties.

Section IV
Ten Most Frequently Reported Major Congenital Malformations by County, 1996-1997
Congenital Malformations Registry

County	ICD-9 Code	Description	Number in 1996	Number in 1997
Albany				
	752.5	Undescended testicle	13	14
	755.0	Polydactyly	13	11
	745.4	Ventricular septal defect	13	9
	752.6	Hypospadias & epispadias	13	8
	754.3	Congenital dislocation of hip	11	15
	745.5	Ostium secundum atrial septal defect	10	20
	754.5	Varus deformities of feet	6	7
	753.2	Obstructive defects of renal pelvis & ureter	6	2
	747.3	Anomalies of pulmonary artery	5	7
	746.0	Anomalies of pulmonary valve	5	5
	750.5	Congenital hypertrophic pyloric stenosis	4	8
	746.8	Other specified anomalies of heart	3	9
Allegany				
	747.3	Anomalies of pulmonary artery	3	3
	752.6	Hypospadias & epispadias	3	1
	754.3	Congenital dislocation of hip	2	1
	745.4	Ventricular septal defect	2	1
	742.4	Other specified anomalies of brain	2	0
	750.5	Congenital hypertrophic pyloric stenosis	2	0
	745.5	Ostium secundum atrial septal defect	1	4
	746.8	Other specified anomalies of heart	1	2
	754.5	Varus deformities of feet	1	2
	756.0	Anomalies of skull and face bones	1	2
	741.0	Spina bifida with hydrocephalus	1	0
	748.0	Choanal atresia	1	0
	743.4	Coloboma & other anomalies of anterior segment	1	0
	749.1	Cleft lip	1	0
	759.5	Tuberous sclerosis	1	0
	755.6	Other anomalies of lower limb including pelvic girdle	1	0
	275.4	Disorders of calcium metabolism	1	0
	758.0	Down syndrome	0	4
	749.0	Cleft palate	0	3
	742.3	Congenital hydrocephalus	0	2
	742.1	Microcephalus	0	2
	343.9	Infantile cerebral palsy unspecified	0	2
	753.2	Obstructive defects of renal pelvis & ureter	0	2
	753.1	Cystic kidney disease	0	2
Bronx				
	755.0	Polydactyly	90	88
	745.4	Ventricular septal defect	84	80
	752.6	Hypospadias & epispadias	74	58
	745.5	Ostium secundum atrial septal defect	68	78
	752.5	Undescended testicle	63	50
	754.5	Varus deformities of feet	61	36
	753.2	Obstructive defects of renal pelvis & ureter	55	43
	754.3	Congenital dislocation of hip	39	28
	750.5	Congenital hypertrophic pyloric stenosis	29	31
	746.8	Other specified anomalies of heart	21	30
Broome				
	745.5	Ostium secundum atrial septal defect	33	18
	745.4	Ventricular septal def ect	14	12
	752.6	Hypospadias & epispadias	9	7

Section IV
Ten Most Frequently Reported Major Congenital Malformations by County, 1996-1997
Congenital Malformations Registry

County	ICD-9 Code	Description	Number in 1996	Number in 1997
Broome (cont.)				
	752.5	Undescended testicle	8	8
	746.8	Other specified anomalies of heart	8	4
	755.0	Polydactyly	7	5
	747.3	Anomalies of pulmonary artery	6	5
	754.5	Varus deformities of feet	6	5
	747.1	Coarctation of aorta	6	3
	750.5	Congenital hypertrophic pyloric stenosis	4	5
	753.2	Obstructive defects of renal pelvis & ureter	4	1
	751.3	Hirschprung's disease & other functional disorders of colon	4	0
	754.3	Congenital dislocation of hip	2	8
Cattaraugus				
	745.4	Ventricular septal defect	10	0
	745.5	Ostium secundum atrial septal defect	8	4
	752.6	Hypospadias & epispadias	8	4
	752.5	Undescended testicle	8	1
	754.5	Varus deformities of feet	5	2
	746.8	Other specified anomalies of heart	4	4
	754.3	Congenital dislocation of hip	3	2
	755.0	Polydactyly	3	1
	750.5	Congenital hypertrophic pyloric stenosis	2	2
	756.7	Anomalies of abdominal wall	2	2
	748.3	Other anomalies of larynx, trachea, & bronchus	2	1
	755.2	Reduction deformities of upper limb	2	0
	742.3	Congenital hydrocephalus	2	0
	746.0	Anomalies of pulmonary valve	2	0
	758.0	Down syndrome	2	0
	755.6	Other anomalies of lower limb including pelvic girdle	1	3
	742.4	Other specified anomalies of brain	0	2
	752.4	Anomalies of cervix, vagina & external female genitalia	0	2
	747.3	Anomalies of pulmonary artery	0	2
	756.0	Anomalies of skull and face bones	0	2
Cayuga				
	752.6	Hypospadias & epispadias	4	2
	750.5	Congenital hypertrophic pyloric stenosis	4	1
	748.3	Other anomalies of larynx, trachea, & bronchus	4	1
	752.5	Undescended testicle	3	4
	745.4	Ventricular septal defect	3	2
	745.5	Ostium secundum atrial septal defect	3	2
	755.6	Other anomalies of lower limb including pelvic girdle	3	0
	753.2	Obstructive defects of renal pelvis & ureter	2	2
	747.1	Coarctation of aorta	2	1
	754.6	Valgus deformities of feet	2	1
	746.8	Other specified anomalies of heart	2	0
	754.3	Congenital dislocation of hip	2	0
	745.1	Transposition of great vessels	2	0
	754.5	Varus deformities of feet	1	3
	753.0	Renal agenesis & dysgenesis	1	2
	754.7	Other deformities of feet	1	2
	753.3	Other specified anomalies of kidney	1	2

Section IV
Ten Most Frequently Reported Major Congenital Malformations by County, 1996-1997
Congenital Malformations Registry

County	ICD-9 Code	Description	Number in 1996	Number in 1997
Chautauqua				
	747.3	Anomalies of pulmonary artery	0	2
	752.6	Hypospadias & epispadias	9	10
	745.5	Ostium secundum atrial septal defect	7	9
	754.5	Varus deformities of feet	6	1
	752.5	Undescended testicle	5	5
	746.8	Other specified anomalies of heart	5	3
	750.5	Congenital hypertrophic pyloric stenosis	4	11
	745.4	Ventricular septal defect	3	9
	752.8	Other specified anomalies of genital organs	3	2
	756.0	Anomalies of skull and face bones	3	2
	755.1	Syndactyly	3	2
	746.0	Anomalies of pulmonary valve	3	1
	754.7	Other deformities of feet	3	1
	751.2	Atresia, stenosis of large intestine, rectum, & anal canal	3	0
	747.3	Anomalies of pulmonary artery	2	6
	277.0	Cystic fibrosis	2	2
	753.2	Obstructive defects of renal pelvis & ureter	2	2
	759.8	Other specified anomalies	2	2
	742.3	Congenital hydrocephalus	2	2
	745.1	Transposition of great vessels	1	3
	524.0	Major anomalies of jaw size	1	2
	758.0	Down syndrome	1	2
	752.7	Indeterminate sex & pseudo-hermaphroditism	0	3
	747.0	Patent ductus arteriosus	0	2
Chemung				
	750.5	Congenital hypertrophic pyloric stenosis	6	3
	752.5	Undescended testicle	5	3
	745.4	Ventricular septal defect	5	1
	752.6	Hypospadias & epispadias	4	5
	753.2	Obstructive defects of renal pelvis & ureter	4	3
	749.0	Cleft palate	4	1
	755.0	Polydactyly	3	1
	742.3	Congenital hydrocephalus	3	0
	754.6	Valgus deformities of feet	3	0
	754.3	Congenital dislocation of hip	2	6
	758.0	Down syndrome	2	2
	745.2	Tetralogy of Fallot	2	2
	754.5	Varus deformities of feet	1	3
	749.2	Cleft palate with cleft lip	1	2
	746.0	Anomalies of pulmonary valve	1	2
	745.5	Ostium secundum atrial septal defect	0	7
	752.8	Other specified anomalies of genital organs	0	2
Chenango				
	745.5	Ostium secundum atrial septal defect	3	1
	753.2	Obstructive defects of renal pelvis & ureter	2	1
	742.3	Congenital hydrocephalus	2	0
	754.3	Congenital dislocation of hip	1	2
	748.3	Other anomalies of larynx, trachea, & bronchus	1	1
	742.4	Other specified anomalies of brain	1	1
	749.2	Cleft palate with cleft lip	1	1
	758.0	Down syndrome	1	1
	750.5	Congenital hypertrophic pyloric stenosis	1	1
	752.5	Undescended testicle	1	1
	755.0	Polydactyly	1	1

Section IV
Ten Most Frequently Reported Major Congenital Malformations by County, 1996-1997
Congenital Malformations Registry

County	ICD-9 Code	Description	Number in 1996	Number in 1997
Chenango (cont.)				
	749.1	Cleft lip	1	1
	755.6	Other anomalies of lower limb including pelvic girdle	1	1
	747.0	Patent ductus arteriosus	1	0
	743.3	Congenital cataract & lens anomalies	1	0
	343.9	Infantile cerebral palsy unspecified	1	0
	743.4	Coloboma & other anomalies of anterior segment	1	0
	745.1	Transposition of great vessels	1	0
	746.8	Other specified anomalies of heart	1	0
	751.1	Atresia & stenosis of small intestine	1	0
	752.6	Hypospadias & epispadias	1	0
	771.2	Other congenital infections	1	0
	756.0	Anomalies of skull and face bones	0	3
	749.0	Cleft palate	0	2
	745.6	Endocardial cushion defects	0	1
	748.1	Other anomalies of nose	0	1
	754.5	Varus deformities of feet	0	1
	754.0	Deformities of skull, face, & jaw	0	1
	753.1	Cystic kidney disease	0	1
	756.8	Other specified anomalies of muscle, tendon, fascia, etc.	0	1
	758.2	Edwards syndrome	0	1
	742.1	Microcephalus	0	1
	752.4	Anomalies of cervix, vagina & external female genitalia	0	1
	751.5	Other anomalies of intestine	0	1
	754.7	Other deformities of feet	0	1
	755.1	Syndactyly	0	1
	756.7	Anomalies of abdominal wall	0	1
	757.3	Other specified anomalies of skin	0	1
	758.1	Patau syndrome	0	1
Clinton				
	753.2	Obstructive defects of renal pelvis & ureter	8	5
	745.4	Ventricular septal defect	7	9
	745.5	Ostium secundum atrial septal defect	4	4
	752.6	Hypospadias & epispadias	3	3
	754.3	Congenital dislocation of hip	2	3
	742.3	Congenital hydrocephalus	2	0
	747.0	Patent ductus arteriosus	1	2
	753.1	Cystic kidney disease	1	2
	752.5	Undescended testicle	1	1
	751.5	Other anomalies of intestine	1	0
	758.7	Klinefelters syndrome	1	0
	277.0	Cystic fibrosis	1	0
	746.0	Anomalies of pulmonary valve	1	0
	750.5	Congenital hypertrophic pyloric stenosis	1	0
	753.0	Renal agenesis & dysgenesis	1	0
	759.5	Tuberous sclerosis	1	0
	749.1	Cleft lip	0	1
	745.2	Tetralogy of Fallot	0	1
	749.0	Cleft palate	0	1
	749.2	Cleft palate with cleft lip	0	1
	752.8	Other specified anomalies of genital organs	0	1
	755.2	Reduction deformities of upper limb	0	1
	756.0	Anomalies of skull and face bones	0	1
	758.0	Down syndrome	0	1

Section IV
Ten Most Frequently Reported Major Congenital Malformations by County, 1996-1997
Congenital Malformations Registry

County	ICD-9 Code	Description	Number in 1996	Number in 1997
Columbia				
	754.3	Congenital dislocation of hip	5	0
	750.5	Congenital hypertrophic pyloric stenosis	4	3
	754.5	Varus deformities of feet	3	1
	745.5	Ostium secundum atrial septal defect	2	4
	745.4	Ventricular septal defect	2	2
	741.9	Spina bifida w/o hydrocephalus	2	1
	741.0	Spina bifida with hydrocephalus	2	0
	747.3	Anomalies of pulmonary artery	2	0
	752.5	Undescended testicle	1	3
	752.6	Hypospadias & epispadias	1	2
	746.0	Anomalies of pulmonary valve	1	1
	742.4	Other specified anomalies of brain	1	1
	742.3	Congenital hydrocephalus	1	1
	746.8	Other specified anomalies of heart	1	1
	748.3	Other anomalies of larynx, trachea, & bronchus	1	0
	746.2	Ebstein's anomaly	1	0
	753.2	Obstructive defects of renal pelvis & ureter	1	0
	745.1	Transposition of great vessels	1	0
	745.3	Common ventricle	1	0
	745.6	Endocardial cushion defects	1	0
	746.3	Congenital stenosis of aortic arch	1	0
	746.4	Congenital insufficiency of aortic arch	1	0
	747.4	Anomalies of great veins	1	0
	751.1	Atresia and stenosis of small intestine	1	0
	753.3	Other specified anomalies of kidney	1	0
	753.7	Anomalies of urachus	1	0
	754.6	Valgus deformities of feet	1	0
	755.6	Other anomalies of lower limb including pelvic girdle	1	0
	756.5	Osteodystrophies	1	0
	759.8	Other specified anomalies	1	0
	751.2	Atresia and stenosis of large intestine, rectum, & anal canal	1	0
	755.2	Reduction deformities of upper limb	0	3
	755.5	Other anomalies of upper limb including shoulder girdle	0	2
	751.6	Anomalies of gallbladder, bile ducts, and liver	0	1
	752.8	Other specified anomalies of genital organs	0	1
	744.8	Other specified anomalies of face & neck	0	1
	747.0	Patent ductus arteriosus	0	1
	742.2	Reduction deformities of brain	0	1
	524.0	Major anomalies of jaw size	0	1
	550.9	Inguinal hernia unspecified	0	1
	743.6	Congenital anomalies of eyelids, lacrimal system and orbit	0	1
	745.2	Tetralogy of Fallot	0	1
	750.2	Other specified anomalies, mouth and pharynx	0	1
	755.1	Syndactyly	0	1
	756.0	Anomalies of skull and face bones	0	1
Cortland				
	745.5	Ostium secundum atrial septal defect	4	1
	747.3	Anomalies of pulmonary artery	3	0
	752.6	Hypospadias & epispadias	2	3
	746.8	Other specified anomalies of heart	2	0
	748.3	Other anomalies of larynx, trachea, & bronchus	1	2
	745.4	Ventricular septal defect	1	2
	750.5	Congenital hypertrophic pyloric stenosis	1	1
	746.0	Anomalies of pulmonary valve	1	1

Section IV
Ten Most Frequently Reported Major Congenital Malformations by County, 1996-1997
Congenital Malformations Registry

County	ICD-9 Code	Description	Number in 1996	Number in 1997
Cortland (cont.)				
	753.2	Obstructive defects of renal pelvis & ureter	1	1
	756.6	Anomalies of diaphragm	1	0
	753.3	Other specified anomalies of kidney	1	0
	756.7	Anomalies of abdominal wall	1	0
	755.0	Polydactyly	1	0
	753.0	Renal agenesis & dysgenesis	1	0
	746.9	Unspecified anomaly of heart	1	0
	755.6	Other anomalies of lower limb including pelvic girdle	1	0
	746.7	Hypoplastic left heart syndrome	1	0
	750.3	Tracheoesophageal fistula, esophageal atresia & stenosis	1	0
	753.1	Cystic kidney disease	1	0
	742.3	Congenital hydrocephalus	0	2
	747.2	Other anomalies of aorta	0	1
	751.5	Other anomalies of intestine	0	1
	749.1	Cleft lip	0	1
	752.5	Undescended testicle	0	1
	754.5	Varus deformities of feet	0	1
	754.8	Other specified nonteratogenic anomalies	0	1
	743.1	Microphthalmos	0	1
	749.0	Cleft palate	0	1
Delaware				
	756.0	Anomalies of skull and face bones	3	4
	752.5	Undescended testicle	2	2
	754.0	Deformities of skull, face, & jaw	2	0
	745.5	Ostium secundum atrial septal defect	1	3
	747.3	Anomalies of pulmonary artery	1	2
	753.0	Renal agenesis & dysgenesis	1	1
	752.6	Hypospadias & epispadias	1	1
	754.3	Congenital dislocation of hip	1	1
	277.0	Cystic fibrosis	1	1
	425.3	Endocardial fibroelastosis	1	0
	742.0	Encephalocele	1	0
	742.3	Congenital hydrocephalus	1	0
	743.3	Congenital cataract & lens anomalies	1	0
	748.3	Other anomalies of larynx, trachea, & bronchus	1	0
	745.4	Ventricular septal defect	1	0
	751.5	Other anomalies of intestine	1	0
	753.5	Exstrophy of urinary bladder	1	0
	753.6	Atresia and stenosis of urethra & bladder neck	1	0
	758.0	Down syndrome	1	0
	759.8	Other specified anomalies	1	0
	753.1	Cystic kidney disease	1	0
	751.6	Anomalies of gallbladder, bile ducts, and liver	1	0
	748.5	Agenesis, hypoplasia & dysplasia, lung	1	0
	756.6	Anomalies of diaphragm	1	0
	748.2	Web of larynx	1	0
	742.2	Reduction deformities of brain	1	0
	742.4	Other specified anomalies of brain	0	2
	750.5	Congenital hypertrophic pyloric stenosis	0	2
	753.2	Obstructive defects of renal pelvis & ureter	0	1
	753.4	Other specified anomalies of ureter	0	1
	746.6	Congenital mitral insufficiency	0	1
	749.0	Cleft palate	0	1
	749.2	Cleft palate with cleft lip	0	1

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County	ICD-9 Code	Description	Number in 1996	Number in 1997
Delaware (cont.)				
	749.1	Cleft lip	0	1
	751.2	Atresia and stenosis of large intestine, rectum, & anal canal	0	1
	755.2	Reduction deformities of upper limb	0	1
Dutchess				
	752.6	Hypospadias & epispadias	19	14
	753.2	Obstructive defects of renal pelvis & ureter	12	9
	752.5	Undescended testicle	9	15
	745.4	Ventricular septal defect	7	14
	755.6	Other anomalies of lower limb including pelvic girdle	6	3
	754.3	Congenital dislocation of hip	5	9
	750.5	Congenital hypertrophic pyloric stenosis	5	7
	745.5	Ostium secundum atrial septal defect	5	6
	755.0	Polydactyly	4	6
	754.5	Varus deformities of feet	4	5
	752.8	Other specified anomalies of genital organs	4	3
	746.8	Other specified anomalies of heart	3	5
	749.0	Cleft palate	2	6
Erie				
	745.5	Ostium secundum atrial septal defect	96	111
	752.5	Undescended testicle	66	78
	745.4	Ventricular septal defect	65	61
	752.6	Hypospadias & epispadias	64	59
	755.0	Polydactyly	58	46
	747.3	Anomalies of pulmonary artery	45	55
	746.8	Other specified anomalies of heart	43	39
	754.5	Varus deformities of feet	27	19
	748.3	Other anomalies of larynx, trachea, & bronchus	25	32
	750.5	Congenital hypertrophic pyloric stenosis	23	29
	753.2	Obstructive defects of renal pelvis & ureter	16	21
Essex				
	753.2	Obstructive defects of renal pelvis & ureter	2	1
	756.5	Osteodystrophies	1	0
	745.4	Ventricular septal defect	1	0
	751.6	Anomalies of gallbladder, bile ducts, and liver	1	0
	752.5	Undescended testicle	1	0
	753.0	Renal agenesis & dysgenesis	1	0
	745.5	Ostium secundum atrial septal defect	0	2
	750.5	Congenital hypertrophic pyloric stenosis	0	1
	752.6	Hypospadias & epispadias	0	1
	754.5	Varus deformities of feet	0	1
	755.2	Reduction deformities of upper limb	0	1
	755.3	Reduction deformities of lower limb	0	1
	756.0	Anomalies of skull and face bones	0	1
	758.0	Down syndrome	0	1
Franklin				
	745.4	Ventricular septal defect	3	1
	745.5	Ostium secundum atrial septal defect	2	1
	753.2	Obstructive defects of renal pelvis & ureter	2	1
	749.0	Cleft palate	2	0
	752.6	Hypospadias & epispadias	1	2
	748.3	Other anomalies of larynx, trachea, & bronchus	1	1
	750.5	Congenital hypertrophic pyloric stenosis	1	1
	752.8	Other specified anomalies of genital organs	1	1
	747.6	Other anomalies of peripheral vascular system	1	0

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County	ICD-9 Code	Description	Number in 1996	Number in 1997
Franklin (cont.)				
	748.5	Agenesis, hypoplasia & dysplasia, lung	1	0
	751.4	Anomalies of intestinal fixation	1	0
	745.2	Tetralogy of Fallot	1	0
	747.3	Anomalies of pulmonary artery	1	0
	754.5	Varus deformities of feet	0	2
	755.0	Polydactyly	0	2
	743.4	Coloboma & other anomalies of anterior segment	0	1
	750.3	Tracheoesophageal fistula, esophageal atresia & stenosis	0	1
	755.1	Syndactyly	0	1
	758.2	Edwards syndrome	0	1
Fulton				
	752.6	Hypospadias & epispadias	5	4
	752.5	Undescended testicle	4	3
	755.0	Polydactyly	4	0
	750.5	Congenital hypertrophic pyloric stenosis	3	4
	748.3	Other anomalies of larynx, trachea, & bronchus	3	2
	745.5	Ostium secundum atrial septal defect	2	2
	747.3	Anomalies of pulmonary artery	2	1
	754.5	Varus deformities of feet	2	1
	746.8	Other specified anomalies of heart	2	1
	754.3	Congenital dislocation of hip	1	3
	745.4	Ventricular septal defect	1	1
	749.1	Cleft lip	1	1
	754.6	Valgus deformities of feet	1	1
	752.0	Anomalies of ovaries	1	0
	746.4	Congenital insufficiency of aortic arch	1	0
	270.1	Phenylketonuria	1	0
	747.0	Patent ductus arteriosus	1	0
	751.4	Anomalies of intestinal fixation	1	0
	751.5	Other anomalies of intestine	1	0
	753.7	Anomalies of urachus	1	0
	755.2	Reduction deformities of upper limb	1	0
	750.2	Other specified anomalies, mouth and pharynx	0	2
	749.2	Cleft palate with cleft lip	0	1
	742.3	Congenital hydrocephalus	0	1
	745.1	Transposition of great vessels	0	1
	746.0	Anomalies of pulmonary valve	0	1
	749.0	Cleft palate	0	1
	755.6	Other anomalies of lower limb including pelvic girdle	0	1
Genesee				
	745.5	Ostium secundum atrial septal defect	5	11
	752.5	Undescended testicle	3	6
	750.5	Congenital hypertrophic pyloric stenosis	3	2
	747.3	Anomalies of pulmonary artery	3	1
	752.6	Hypospadias & epispadias	3	0
	753.2	Obstructive defects of renal pelvis & ureter	3	0
	746.8	Other specified anomalies of heart	2	3
	752.7	Indeterminate sex & pseudo-hermaphroditism	2	0
	756.0	Anomalies of skull and face bones	2	0
	745.4	Ventricular septal defect	1	2
	758.0	Down syndrome	1	2
	746.0	Anomalies of pulmonary valve	1	1
	754.3	Congenital dislocation of hip	1	1
	748.5	Agenesis, hypoplasia & dysplasia, lung	1	1

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County	ICD-9 Code	Description	Number in 1996	Number in 1997
Genesee (cont.)				
	749.0	Cleft palate	1	1
	756.6	Anomalies of diaphragm	1	1
	758.6	Gonadal dysgenesis	1	1
	753.0	Renal agenesis & dysgenesis	1	0
	749.2	Cleft palate with cleft lip	1	0
	745.2	descripetralogy of Fallot	1	0
	273.8	Other disorders of plasma protien	1	0
	743.3	Congenital cataract & lens anomalies	1	0
	746.3	Congenital stenosis of aortic arch	1	0
	748.0	Choanal atresia	1	0
	748.4	Congenital cystic lung	1	0
	751.4	Anomalies of intestinal fixation	1	0
	751.5	Other anomalies of intestine	1	0
	751.6	Anomalies of gallbladder, bile ducts, and liver	1	0
	753.1	Cystic kidney disease	1	0
	754.5	Varus deformities of feet	1	0
	754.7	Other deformities of feet	1	0
	758.8	Other conditions due to sex chromosome anomalies	1	0
	759.0	Anomalies of spleen	1	0
	759.8	Other specified anomalies	1	0
	755.6	Other anomalies of lower limb including pelvic girdle	0	2
	243.	Congenital hypothyroidism	0	1
	277.0	Cystic fibrosis	0	1
	282.0	Hereditary spherocytosis	0	1
	740.0	Anencephalus	0	1
	744.0	Anomalies of ear causing impairment of hearing	0	1
	747.0	Patent ductus arteriosus	0	1
	750.3	descripracheoesophageal fistula, esophageal atresia & stenosis	0	1
	751.1	Atresia and stenosis of small intestine	0	1
	751.3	Hirschprung's disease & other functional disorders of colon	0	1
	752.8	Other specified anomalies of genital organs	0	1
	754.6	Valgus deformities of feet	0	1
	755.0	Polydactyly	0	1
	756.5	Osteodystrophies	0	1
Greene				
	754.3	Congenital dislocation of hip	3	2
	745.4	Ventricular septal defect	2	4
	742.2	Reduction deformities of brain	2	0
	747.1	Coarctation of aorta	1	4
	750.5	Congenital hypertrophic pyloric stenosis	1	1
	754.6	Valgus deformities of feet	1	1
	741.0	Spina bifida with hydrocephalus	1	0
	751.3	Hirschprung's disease & other functional disorders of colon	1	0
	752.5	Undescended testicle	1	0
	742.3	Congenital hydrocephalus	1	0
	755.6	Other anomalies of lower limb including pelvic girdle	1	0
	756.1	Anomalies of spine	1	0
	752.6	Hypospadias & epispadias	0	3
	746.3	Congenital stenosis of aortic arch	0	2
	752.8	Other specified anomalies of genital organs	0	1
	748.5	Agenesis, hypoplasia & dysplasia, lung	0	1
	746.4	Congenital insufficiency of aortic arch	0	1
	746.6	Congenital mitral insufficiency	0	1

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County	ICD-9 Code	Description	Number in 1996	Number in 1997
Greene (cont.)				
	746.7	Hypoplastic left heart syndrome	0	1
	746.8	Other specified anomalies of heart	0	1
	759.2	Anomalies of other endocrine glands	0	1
	749.0	Cleft palate	0	1
	753.2	Obstructive defects of renal pelvis & ureter	0	1
	754.5	Varus deformities of feet	0	1
	745.5	Ostium secundum atrial septal defect	0	1
	746.5	Congenital mitral stenosis	0	1
Hamilton				
	749.2	Cleft palate with cleft lip	1	0
	750.5	Congenital hypertrophic pyloric stenosis	1	0
	749.1	Cleft lip	1	0
Herkimer				
	754.3	Congenital dislocation of hip	4	3
	752.6	Hypospadias & epispadias	3	4
	745.4	Ventricular septal defect	3	4
	754.5	Varus deformities of feet	3	2
	753.2	Obstructive defects of renal pelvis & ureter	3	1
	755.6	Other anomalies of lower limb including pelvic girdle	2	5
	752.5	Undescended testicle	2	3
	742.4	Other specified anomalies of brain	1	3
	750.2	Other specified anomalies, mouth and pharynx	1	2
	750.5	Congenital hypertrophic pyloric stenosis	1	2
	756.7	Anomalies of abdominal wall	1	1
	751.4	Anomalies of intestinal fixation	1	0
	524.0	Major anomalies of jaw size	1	0
	745.5	Ostium secundum atrial septal defect	1	0
	752.8	Other specified anomalies of genital organs	1	0
	745.0	Common truncus	1	0
	742.1	Microcephalus	1	0
	758.0	Down syndrome	1	0
	751.5	Other anomalies of intestine	1	0
	746.0	Anomalies of pulmonary valve	1	0
	742.3	Congenital hydrocephalus	0	2
	755.0	Polydactyly	0	2
	754.8	Other specified nonteratogenic anomalies	0	2
Jefferson				
	745.4	Ventricular septal defect	9	8
	752.6	Hypospadias & epispadias	8	2
	747.3	Anomalies of pulmonary artery	6	0
	752.5	Undescended testicle	5	4
	750.5	Congenital hypertrophic pyloric stenosis	5	3
	745.5	Ostium secundum atrial septal defect	5	0
	754.3	Congenital dislocation of hip	3	3
	754.5	Varus deformities of feet	3	3
	742.3	Congenital hydrocephalus	3	2
	753.2	Obstructive defects of renal pelvis & ureter	2	3
	749.2	Cleft palate with cleft lip	2	2
	749.0	Cleft palate	2	1
	750.3	Tracheoesophageal fistula, esophageal atresia & stenosis	2	0
	756.0	Anomalies of skull and face bones	2	0
	758.0	Down syndrome	2	0
	748.3	Other anomalies of larynx, trachea, & bronchus	1	5
	755.0	Polydactyly	1	5
	756.7	Anomalies of abdominal wall	1	2

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County	ICD-9 Code	Description	Number in 1996	Number in 1997
Jefferson (cont.)				
	751.2	Atresia and stenosis of large intestine, rectum, & anal canal	0	2
	751.1	Atresia and stenosis of small intestine	0	2
	755.1	Syndactyly	0	2
	755.2	Reduction deformities of upper limb	0	2
	756.1	Anomalies of spine	0	2
	759.8	Other specified anomalies	0	2
Kings				
	745.5	Ostium secundum atrial septal defect	342	320
	745.4	Ventricular septal defect	144	152
	755.0	Polydactyly	138	118
	752.6	Hypospadias & epispadias	110	90
	753.2	Obstructive defects of renal pelvis & ureter	89	84
	754.5	Varus deformities of feet	84	68
	752.5	Undescended testicle	77	88
	746.8	Other specified anomalies of heart	76	51
	746.0	Anomalies of pulmonary valve	55	38
	750.5	Congenital hypertrophic pyloric stenosis	54	53
Lewis				
	745.4	Ventricular septal defect	3	1
	750.5	Congenital hypertrophic pyloric stenosis	3	1
	754.3	Congenital dislocation of hip	2	3
	753.2	Obstructive defects of renal pelvis & ureter	2	1
	756.0	Anomalies of skull and face bones	2	1
	752.5	Undescended testicle	2	1
	755.6	Other anomalies of lower limb including pelvic girdle	2	0
	752.6	Hypospadias & epispadias	1	3
	749.0	Cleft palate	1	2
	754.5	Varus deformities of feet	1	2
	745.5	Ostium secundum atrial septal defect	1	1
	758.0	Down syndrome	1	1
	748.1	Other anomalies of nose	1	1
	758.7	Klinefelters syndrome	1	0
	747.1	Coarctation of aorta	1	0
	448.0	Hereditary hemorrhagic telangiectasia	1	0
	742.3	Congenital hydrocephalus	1	0
	747.3	Anomalies of pulmonary artery	1	0
	750.2	Other specified anomalies, mouth and pharynx	1	0
	751.3	Hirschprung's disease & other functional disorders of colon	1	0
	752.8	Other specified anomalies of genital organs	1	0
	755.0	Polydactyly	1	0
	755.1	Syndactyly	1	0
	753.4	Other specified anomalies of ureter	1	0
	741.9	Spina bifida w/o hydrocephalus	0	1
	742.1	Microcephalus	0	1
	742.2	Reduction deformities of brain	0	1
	742.4	Other specified anomalies of brain	0	1
	747.0	Patent ductus arteriosus	0	1
	749.2	Cleft palate with cleft lip	0	1
	758.2	Edwards syndrome	0	1
Livingston				
	752.6	Hypospadias & epispadias	4	2
	745.4	Ventricular septal defect	3	0
	750.5	Congenital hypertrophic pyloric stenosis	2	2
	746.8	Other specified anomalies of heart	2	1
	747.2	Other anomalies of aorta	2	0

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County	ICD-9 Code	Description	Number in 1996	Number in 1997
Livingston (cont.)				
	747.1	Coarctation of aorta	2	0
	754.3	Congenital dislocation of hip	1	3
	755.0	Polydactyly	1	1
	751.1	Atresia and stenosis of small intestine	1	0
	755.1	Syndactyly	1	0
	745.5	Ostium secundum atrial septal defect	1	0
	275.4	Disorders of calcium metabolism	1	0
	228.1	Lymphangioma, any site	1	0
	758.7	Klinefelters syndrome	1	0
	746.5	Congenital mitral stenosis	1	0
	746.7	Hypoplastic left heart syndrome	1	0
	753.2	Obstructive defects of renal pelvis & ureter	1	0
	754.5	Varus deformities of feet	0	3
	752.5	Undescended testicle	0	2
	754.6	Valgus deformities of feet	0	2
	747.3	Anomalies of pulmonary artery	0	1
	426.7	Anamalous atrioventricular excitation	0	1
	742.5	Other specified anomalies of spinal cord	0	1
	743.1	Microphthalmos	0	1
	743.3	Congenital cataract & lens anomalies	0	1
	743.6	Congenital anomalies of eyelids, lacrimal system and orbit	0	1
	746.0	Anomalies of pulmonary valve	0	1
	746.6	Congenital mitral insufficiency	0	1
	748.3	Other anomalies of larynx, trachea, & bronchus	0	1
	751.5	Other anomalies of intestine	0	1
	759.8	Other specified anomalies	0	1
	754.0	Deformities of skull, face, & jaw	0	1
	756.7	Anomalies of abdominal wall	0	1
	758.1	Patau syndrome	0	1
	756.3	Other anomalies of ribs and sternum	0	1
	756.0	Anomalies of skull and face bones	0	1
	757.3	Other specified anomalies of skin	0	1
Madison				
	752.5	Undescended testicle	5	4
	750.5	Congenital hypertrophic pyloric stenosis	4	4
	755.0	Polydactyly	4	2
	754.5	Varus deformities of feet	3	3
	752.6	Hypospadias & epispadias	3	1
	745.5	Ostium secundum atrial septal defect	2	4
	753.2	Obstructive defects of renal pelvis & ureter	2	1
	746.8	Other specified anomalies of heart	2	1
	755.5	Other anomalies of upper limb including shoulder girdle	2	0
	742.1	Microcephalus	2	0
	745.4	Ventricular septal defect	1	4
	746.0	Anomalies of pulmonary valve	1	2
	749.0	Cleft palate	1	2
	748.3	Other anomalies of larynx, trachea, & bronchus	0	3
	751.2	Atresia and stenosis of large intestine, rectum, & anal canal	0	2
	756.0	Anomalies of skull and face bones	0	2
Monroe				
	752.6	Hypospadias & epispadias	43	31
	752.5	Undescended testicle	26	29
	745.4	Ventricular septal defect	20	24
	750.5	Congenital hypertrophic pyloric stenosis	20	21
	755.0	Polydactyly	18	27

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Monroe (cont.)				
	753.2	Obstructive defects of renal pelvis & ureter	18	24
	754.5	Varus deformities of feet	18	20
	746.8	Other specified anomalies of heart	14	13
	754.3	Congenital dislocation of hip	14	10
	746.0	Anomalies of pulmonary valve	13	5
	758.0	Down syndrome	10	16
	745.5	Ostium secundum atrial septal defect	7	17
Montgomery				
	750.5	Congenital hypertrophic pyloric stenosis	5	3
	754.5	Varus deformities of feet	5	2
	752.6	Hypospadias & epispadias	3	0
	755.6	Other anomalies of lower limb including pelvic girdle	3	0
	745.4	Ventricular septal defect	2	2
	745.5	Ostium secundum atrial septal defect	2	2
	744.0	Anomalies of ear causing impairment of hearing	2	0
	746.8	Other specified anomalies of heart	1	2
	754.3	Congenital dislocation of hip	1	2
	756.7	Anomalies of abdominal wall	1	1
	748.5	Agenesis, hypoplasia & dysplasia, lung	1	1
	753.2	Obstructive defects of renal pelvis & ureter	1	1
	752.4	Anomalies of cervix, vagina & external female genitalia	1	1
	759.6	Other hamartoses,nec	1	0
	743.6	Congenital anomalies of eyelids, lacrimal system and orbit	1	0
	752.8	Other specified anomalies of genital organs	1	0
	743.8	Other specified anomalies of eye	1	0
	756.6	Anomalies of diaphragm	1	0
	756.0	Anomalies of skull and face bones	1	0
	751.5	Other anomalies of intestine	1	0
	745.1	Transposition of great vessels	1	0
	747.1	Coarctation of aorta	1	0
	744.3	Unspecified anomaly of ear	1	0
	748.3	Other anomalies of larynx, trachea, & bronchus	0	3
	749.2	Cleft palate with cleft lip	0	2
	752.5	Undescended testicle	0	2
	746.0	Anomalies of pulmonary valve	0	2
Nassau				
	752.6	Hypospadias & epispadias	96	110
	745.4	Ventricular septal defect	88	87
	745.5	Ostium secundum atrial septal defect	84	89
	752.5	Undescended testicle	69	79
	753.2	Obstructive defects of renal pelvis & ureter	69	65
	754.5	Varus deformities of feet	35	41
	755.0	Polydactyly	33	34
	754.3	Congenital dislocation of hip	31	38
	746.8	Other specified anomalies of heart	29	28
	750.5	Congenital hypertrophic pyloric stenosis	28	42
New York				
	745.4	Ventricular septal defect	85	69
	745.5	Ostium secundum atrial septal defect	70	79
	754.3	Congenital dislocation of hip	52	56
	752.6	Hypospadias & epispadias	42	60
	753.2	Obstructive defects of renal pelvis & ureter	41	33
	755.0	Polydactyly	39	39
	746.0	Anomalies of pulmonary valve	30	28
	754.5	Varus deformities of feet	29	21

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County	ICD-9 Code	Description	Number in 1996	Number in 1997
New York (cont.)				
	752.5	Undescended testicle	20	30
	742.3	Congenital hydrocephalus	20	7
Niagara				
	745.4	Ventricular septal defect	16	12
	752.5	Undescended testicle	15	12
	745.5	Ostium secundum atrial septal defect	14	18
	746.8	Other specified anomalies of heart	12	7
	752.6	Hypospadias & epispadias	11	8
	747.3	Anomalies of pulmonary artery	11	6
	755.0	Polydactyly	10	10
	754.5	Varus deformities of feet	7	5
	748.3	Other anomalies of larynx, trachea, & bronchus	6	2
	750.5	Congenital hypertrophic pyloric stenosis	5	6
	753.2	Obstructive defects of renal pelvis & ureter	4	7
	754.3	Congenital dislocation of hip	3	5
	746.0	Anomalies of pulmonary valve	3	5
	742.3	Congenital hydrocephalus	2	5
Oneida				
	752.6	Hypospadias & epispadias	12	11
	754.3	Congenital dislocation of hip	11	10
	745.4	Ventricular septal defect	11	7
	753.2	Obstructive defects of renal pelvis & ureter	11	7
	750.5	Congenital hypertrophic pyloric stenosis	9	10
	747.3	Anomalies of pulmonary artery	9	5
	754.5	Varus deformities of feet	8	7
	752.5	Undescended testicle	6	12
	745.5	Ostium secundum atrial septal defect	5	9
	748.3	Other anomalies of larynx, trachea, & bronchus	5	3
	755.0	Polydactyly	3	5
Onondaga				
	745.4	Ventricular septal defect	32	21
	745.5	Ostium secundum atrial septal defect	31	23
	752.6	Hypospadias & epispadias	31	16
	752.5	Undescended testicle	18	15
	755.0	Polydactyly	17	15
	754.5	Varus deformities of feet	16	11
	747.3	Anomalies of pulmonary artery	16	8
	750.5	Congenital hypertrophic pyloric stenosis	15	10
	746.8	Other specified anomalies of heart	13	15
	753.2	Obstructive defects of renal pelvis & ureter	13	8
	748.3	Other anomalies of larynx, trachea, & bronchus	11	14
	754.3	Congenital dislocation of hip	10	8
Ontario				
	752.6	Hypospadias & epispadias	12	7
	758.0	Down syndrome	7	1
	752.5	Undescended testicle	5	3
	745.4	Ventricular septal defect	4	5
	755.0	Polydactyly	4	1
	754.3	Congenital dislocation of hip	3	7
	753.2	Obstructive defects of renal pelvis & ureter	3	5
	750.5	Congenital hypertrophic pyloric stenosis	3	3
	749.0	Cleft palate	1	4
	746.0	Anomalies of pulmonary valve	1	4
	752.8	Other specified anomalies of genital organs	1	4

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County	ICD-9 Code	Description	Number in 1996	Number in 1997
Ontario (cont.)				
	756.0	Anomalies of skull and face bones	1	3
	755.6	Other anomalies of lower limb including pelvic girdle	1	2
	747.3	Anomalies of pulmonary artery	1	2
	755.5	Other anomalies of upper limb including shoulder girdle	1	2
	749.1	Cleft lip	1	1
	745.5	Ostium secundum atrial septal defect	1	1
	749.2	Cleft palate with cleft lip	1	1
	751.3	Hirschprung's disease & other functional disorders of colon	1	0
	746.7	Hypoplastic left heart syndrome	1	0
	756.7	Anomalies of abdominal wall	1	0
	748.5	Agenesis, hypoplasia & dysplasia, lung	1	0
	741.0	Spina bifida with hydrocephalus	1	0
	748.4	Congenital cystic lung	1	0
	750.7	Other specified anomalies of stomach	1	0
	755.1	Syndactyly	1	0
	745.6	Endocardial cushion defects	1	0
	754.5	Varus deformities of feet	0	5
Orange				
	745.4	Ventricular septal defect	18	20
	752.6	Hypospadias & epispadias	17	24
	752.5	Undescended testicle	17	14
	753.2	Obstructive defects of renal pelvis & ureter	9	8
	754.3	Congenital dislocation of hip	9	5
	754.5	Varus deformities of feet	9	4
	745.5	Ostium secundum atrial septal defect	8	10
	755.0	Polydactyly	7	6
	758.0	Down syndrome	7	6
	756.0	Anomalies of skull and face bones	6	1
	750.5	Congenital hypertrophic pyloric stenosis	5	10
	748.3	Other anomalies of larynx, trachea, & bronchus	3	6
	747.1	Coarctation of aorta	0	7
Orleans				
	746.8	Other specified anomalies of heart	4	0
	745.5	Ostium secundum atrial septal defect	3	1
	752.6	Hypospadias & epispadias	3	1
	754.3	Congenital dislocation of hip	3	0
	750.5	Congenital hypertrophic pyloric stenosis	2	2
	755.0	Polydactyly	2	2
	754.5	Varus deformities of feet	2	0
	755.2	Reduction deformities of upper limb	2	0
	749.1	Cleft lip	2	0
	747.3	Anomalies of pulmonary artery	1	2
	747.4	Anomalies of great veins	1	1
	753.2	Obstructive defects of renal pelvis & ureter	1	1
	749.2	Cleft palate with cleft lip	1	0
	752.8	Other specified anomalies of genital organs	1	0
	745.6	Endocardial cushion defects	1	0
	745.2	Tetralogy of Fallot	1	0
	746.7	Hypoplastic left heart syndrome	1	0
	755.1	Syndactyly	1	0
	524.0	Major anomalies of jaw size	1	0
	745.4	Ventricular septal defect	0	4
	743.6	Congenital anomalies of eyelids, lacrimal system and orbit	0	2
	744.0	Anomalies of ear causing impairment of hearing	0	1

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County	ICD-9 Code	Description	Number in 1996	Number in 1997
Orleans (cont.)				
	742.3	Congenital hydrocephalus	0	1
	742.4	Other specified anomalies of brain	0	1
	743.5	Congenital anomalies of posterior segment of eye	0	1
	745.0	Common truncus	0	1
	748.0	Choanal atresia	0	1
	746.6	Congenital mitral insufficiency	0	1
	752.5	Undescended testicle	0	1
	758.3	Autosomal deletion syndromes	0	1
	748.3	Other anomalies of larynx, trachea, & bronchus	0	1
	754.8	Other specified nonteratogenic anomalies	0	1
	748.5	Agenesis, hypoplasia & dysplasia, lung	0	1
	756.0	Anomalies of skull and face bones	0	1
	756.1	Anomalies of spine	0	1
	756.7	Anomalies of abdominal wall	0	1
	758.0	Down syndrome	0	1
Oswego				
	750.5	Congenital hypertrophic pyloric stenosis	10	5
	752.6	Hypospadias & epispadias	7	8
	753.2	Obstructive defects of renal pelvis & ureter	7	5
	754.3	Congenital dislocation of hip	7	2
	745.4	Ventricular septal defect	6	7
	752.5	Undescended testicle	6	6
	745.5	Ostium secundum atrial septal defect	6	4
	747.3	Anomalies of pulmonary artery	5	4
	749.0	Cleft palate	4	2
	749.2	Cleft palate with cleft lip	4	1
	755.0	Polydactyly	1	3
	748.3	Other anomalies of larynx, trachea, & bronchus	1	2
	751.1	Atresia and stenosis of small intestine	1	2
	754.5	Varus deformities of feet	1	2
	756.0	Anomalies of skull and face bones	1	2
	742.4	Other specified anomalies of brain	0	2
Otsego				
	752.6	Hypospadias & epispadias	2	2
	750.5	Congenital hypertrophic pyloric stenosis	2	0
	752.8	Other specified anomalies of genital organs	2	0
	745.5	Ostium secundum atrial septal defect	1	2
	754.5	Varus deformities of feet	1	1
	741.9	Spina bifida w/o hydrocephalus	1	1
	745.4	Ventricular septal defect	1	1
	758.0	Down syndrome	1	1
	755.0	Polydactyly	1	1
	749.2	Cleft palate with cleft lip	1	0
	750.3	Tracheoesophageal fistula, esophageal atresia & stenosis	1	0
	755.6	Other anomalies of lower limb including pelvic girdle	1	0
	756.6	Anomalies of diaphragm	1	0
	752.0	Anomalies of ovaries	1	0
	746.0	Anomalies of pulmonary valve	1	0
	286.0	Congenital factor VIII disorder	1	0
	746.3	Congenital stenosis of aortic arch	1	0
	748.3	Other anomalies of larynx, trachea, & bronchus	1	0
	753.2	Obstructive defects of renal pelvis & ureter	0	4
	741.0	Spina bifida with hydrocephalus	0	2
	742.3	Congenital hydrocephalus	0	1

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County	ICD-9 Code	Description	Number in 1996	Number in 1997
Otsego (cont.)				
	745.2	Tetralogy of Fallot	0	1
	745.6	Endocardial cushion defects	0	1
	746.8	Other specified anomalies of heart	0	1
	747.0	Patent ductus arteriosus	0	1
	749.1	Cleft lip	0	1
	752.5	Undescended testicle	0	1
	753.1	Cystic kidney disease	0	1
	758.5	Other conditions due autosomal anomalies	0	1
Putnam				
	752.5	Undescended testicle	7	5
	752.6	Hypospadias & epispadias	5	14
	753.2	Obstructive defects of renal pelvis & ureter	3	4
	745.4	Ventricular septal defect	2	3
	742.3	Congenital hydrocephalus	2	0
	754.5	Varus deformities of feet	2	0
	754.3	Congenital dislocation of hip	1	2
	758.0	Down syndrome	1	2
	746.8	Other specified anomalies of heart	1	2
	749.2	Cleft palate with cleft lip	1	1
	750.5	Congenital hypertrophic pyloric stenosis	1	1
	751.1	Atresia and stenosis of small intestine	1	1
	755.1	Syndactyly	1	1
	748.3	Other anomalies of larynx, trachea, & bronchus	1	1
	755.3	Reduction deformities of lower limb	1	0
	756.7	Anomalies of abdominal wall	1	0
	749.1	Cleft lip	1	0
	553.1	Umbilical hernia	1	0
	243.	Congenital hypothyroidism	1	0
	251.2	Hypoglycemia unspecified	1	0
	742.4	Other specified anomalies of brain	1	0
	743.3	Congenital cataract & lens anomalies	1	0
	748.1	Other anomalies of nose	1	0
	754.6	Valgus deformities of feet	1	0
	745.5	Ostium secundum atrial septal defect	0	5
	746.0	Anomalies of pulmonary valve	0	3
	749.0	Cleft palate	0	2
	752.8	Other specified anomalies of genital organs	0	2
	753.6	Atresia and stenosis of urethra & bladder neck	0	2
Queens				
	745.5	Ostium secundum atrial septal defect	136	146
	745.4	Ventricular septal defect	112	120
	752.6	Hypospadias & epispadias	102	123
	755.0	Polydactyly	94	92
	752.5	Undescended testicle	87	90
	753.2	Obstructive defects of renal pelvis & ureter	77	60
	754.3	Congenital dislocation of hip	69	59
	754.5	Varus deformities of feet	45	48
	746.8	Other specified anomalies of heart	40	43
	750.5	Congenital hypertrophic pyloric stenosis	38	43
Rensselaer				
	750.5	Congenital hypertrophic pyloric stenosis	6	3
	754.3	Congenital dislocation of hip	5	10
	745.5	Ostium secundum atrial septal defect	5	5
	755.0	Polydactyly	5	5

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County	ICD-9 Code	Description	Number in 1996	Number in 1997
Rensselaer (cont.)				
	754.5	Varus deformities of feet	5	4
	752.6	Hypospadias & epispadias	4	6
	745.4	Ventricular septal defect	4	3
	752.5	Undescended testicle	3	3
	753.2	Obstructive defects of renal pelvis & ureter	3	3
	748.3	Other anomalies of larynx, trachea, & bronchus	3	2
	746.0	Anomalies of pulmonary valve	3	2
	749.1	Cleft lip	3	1
	756.7	Anomalies of abdominal wall	3	0
	742.1	Microcephalus	3	0
	742.2	Reduction deformities of brain	3	0
	755.6	Other anomalies of lower limb including pelvic girdle	1	3
Richmond				
	752.6	Hypospadias & epispadias	30	28
	745.5	Ostium secundum atrial septal defect	29	42
	745.4	Ventricular septal defect	22	35
	752.5	Undescended testicle	16	13
	753.2	Obstructive defects of renal pelvis & ureter	15	18
	754.5	Varus deformities of feet	13	17
	755.0	Polydactyly	11	11
	746.8	Other specified anomalies of heart	11	9
	755.6	Other anomalies of lower limb including pelvic girdle	9	3
	746.0	Anomalies of pulmonary valve	7	7
	755.1	Syndactyly	7	4
	758.0	Down syndrome	6	9
	750.5	Congenital hypertrophic pyloric stenosis	4	8
Rockland				
	752.6	Hypospadias & epispadias	25	15
	745.4	Ventricular septal defect	14	12
	752.5	Undescended testicle	13	8
	745.5	Ostium secundum atrial septal defect	11	11
	755.0	Polydactyly	10	9
	753.2	Obstructive defects of renal pelvis & ureter	8	6
	754.5	Varus deformities of feet	8	6
	742.3	Congenital hydrocephalus	8	1
	758.0	Down syndrome	5	6
	752.8	Other specified anomalies of genital organs	5	4
	753.1	Cystic kidney disease	5	1
	750.5	Congenital hypertrophic pyloric stenosis	4	9
	755.6	Other anomalies of lower limb including pelvic girdle	1	6
Saratoga				
	752.6	Hypospadias & epispadias	15	11
	754.3	Congenital dislocation of hip	9	11
	745.4	Ventricular septal defect	9	5
	752.5	Undescended testicle	8	8
	750.5	Congenital hypertrophic pyloric stenosis	6	11
	754.5	Varus deformities of feet	6	8
	746.4	Congenital insufficiency of aortic arch	4	1
	749.2	Cleft palate with cleft lip	4	1
	758.0	Down syndrome	3	3
	756.0	Anomalies of skull and face bones	3	3
	745.5	Ostium secundum atrial septal defect	3	2
	753.2	Obstructive defects of renal pelvis & ureter	3	1
	745.1	Transposition of great vessels	3	0

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County	ICD-9 Code	Description	Number in 1996	Number in 1997
Saratoga (cont.)				
	742.4	Other specified anomalies of brain	2	4
	748.3	Other anomalies of larynx, trachea, & bronchus	2	3
	755.0	Polydactyly	1	5
Schenectady				
	752.6	Hypospadias & epispadias	8	11
	745.4	Ventricular septal defect	8	1
	745.5	Ostium secundum atrial septal defect	7	2
	753.2	Obstructive defects of renal pelvis & ureter	6	3
	754.5	Varus deformities of feet	5	5
	750.5	Congenital hypertrophic pyloric stenosis	5	3
	752.5	Undescended testicle	4	7
	749.2	Cleft palate with cleft lip	4	2
	756.0	Anomalies of skull and face bones	4	0
	755.0	Polydactyly	3	5
	749.0	Cleft palate	3	0
	746.8	Other specified anomalies of heart	3	0
	742.3	Congenital hydrocephalus	2	3
	746.0	Anomalies of pulmonary valve	2	2
	754.3	Congenital dislocation of hip	1	5
	756.3	Other anomalies of ribs and sternum	0	2
	754.6	Valgus deformities of feet	0	2
Schoharie				
	745.5	Ostium secundum atrial septal defect	3	2
	746.8	Other specified anomalies of heart	2	0
	752.5	Undescended testicle	2	0
	752.6	Hypospadias & epispadias	1	3
	745.4	Ventricular septal defect	1	1
	758.0	Down syndrome	1	1
	747.6	Other anomalies of peripheral vascular system	1	0
	747.2	Other anomalies of aorta	1	0
	756.6	Anomalies of diaphragm	1	0
	756.5	Osteodystrophies	1	0
	749.0	Cleft palate	1	0
	747.3	Anomalies of pulmonary artery	1	0
	750.5	Congenital hypertrophic pyloric stenosis	1	0
	742.4	Other specified anomalies of brain	1	0
	748.2	Web of larynx	1	0
	746.1	Tricuspid atresia & stenosis	1	0
	745.0	Common truncus	1	0
	754.3	Congenital dislocation of hip	0	2
	755.0	Polydactyly	0	1
	748.0	Choanal atresia	0	1
	753.1	Cystic kidney disease	0	1
	754.6	Valgus deformities of feet	0	1
	742.1	Microcephalus	0	1
	742.3	Congenital hydrocephalus	0	1
	741.9	Spina bifida w/o hydrocephalus	0	1
	743.4	Coloboma & other anomalies of anterior segment	0	1
	745.1	Transposition of great vessels	0	1
Schuyler				
	741.0	Spina bifida with hydrocephalus	2	0
	747.0	Patent ductus arteriosus	2	0
	745.5	Ostium secundum atrial septal defect	1	0
	748.3	Other anomalies of larynx, trachea, & bronchus	1	0

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County	ICD-9 Code	Description	Number in 1996	Number in 1997
Schuyler (cont.)				
	751.3	Hirschprung's disease & other functional disorders of colon	1	0
	524.0	Major anomalies of jaw size	1	0
	741.9	Spina bifida w/o hydrocephalus	1	0
	742.2	Reduction deformities of brain	1	0
	742.0	Encephalocele	1	0
	746.8	Other specified anomalies of heart	1	0
	749.0	Cleft palate	1	0
	750.3	Tracheoesophageal fistula, esophageal atresia & stenosis	1	0
	754.3	Congenital dislocation of hip	1	0
	754.5	Varus deformities of feet	1	0
	756.7	Anomalies of abdominal wall	1	0
	759.8	Other specified anomalies	1	0
	752.5	Undescended testicle	1	0
	742.3	Congenital hydrocephalus	1	0
	754.6	Valgus deformities of feet	1	0
	755.0	Polydactyly	0	2
	752.6	Hypospadias & epispadias	0	1
	747.1	Coarctation of aorta	0	1
	752.8	Other specified anomalies of genital organs	0	1
	745.4	Ventricular septal defect	0	1
	745.1	Transposition of great vessels	0	1
	746.7	Hypoplastic left heart syndrome	0	1
Seneca				
	752.5	Undescended testicle	2	2
	747.0	Patent ductus arteriosus	2	1
	750.5	Congenital hypertrophic pyloric stenosis	2	1
	749.0	Cleft palate	2	0
	745.4	Ventricular septal defect	2	0
	754.0	Deformities of skull, face, & jaw	1	1
	752.6	Hypospadias & epispadias	1	1
	753.2	Obstructive defects of renal pelvis & ureter	1	1
	745.5	Ostium secundum atrial septal defect	1	0
	270.2	Other disturbances of aromatic amino acid metabolism	1	0
	742.1	Microcephalus	1	0
	745.0	Common truncus	1	0
	746.8	Other specified anomalies of heart	1	0
	747.3	Anomalies of pulmonary artery	1	0
	749.2	Cleft palate with cleft lip	1	0
	752.8	Other specified anomalies of genital organs	1	0
	753.1	Cystic kidney disease	1	0
	756.0	Anomalies of skull and face bones	1	0
	754.4	Congenital genu recurvatum & bowing of long bones of leg	1	0
	742.3	Congenital hydrocephalus	1	0
	754.6	Valgus deformities of feet	1	0
	752.0	Anomalies of ovaries	1	0
	754.5	Varus deformities of feet	0	2
	755.3	Reduction deformities of lower limb	0	1
	755.6	Other anomalies of lower limb including pelvic girdle	0	1
	756.4	Chondrodystrophy	0	1
	743.3	Congenital cataract & lens anomalies	0	1
	743.4	Coloboma & other anomalies of anterior segment	0	1
	746.9	Unspecified anomaly of heart	0	1
	754.3	Congenital dislocation of hip	0	1
	755.2	Reduction deformities of upper limb	0	1

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County	ICD-9 Code	Description	Number in 1996	Number in 1997
St. Lawrence				
	752.6	Hypospadias & epispadias	8	4
	750.5	Congenital hypertrophic pyloric stenosis	7	0
	745.4	Ventricular septal defect	6	4
	754.5	Varus deformities of feet	5	3
	745.5	Ostium secundum atrial septal defect	4	3
	752.5	Undescended testicle	4	2
	742.3	Congenital hydrocephalus	3	1
	754.6	Valgus deformities of feet	3	0
	756.0	Anomalies of skull and face bones	3	0
	754.0	Deformities of skull, face, & jaw	2	1
	758.0	Down syndrome	2	1
	753.1	Cystic kidney disease	2	1
	746.8	Other specified anomalies of heart	2	0
	749.0	Cleft palate	2	0
	749.1	Cleft lip	2	0
	749.2	Cleft palate with cleft lip	2	0
	755.1	Syndactyly	1	2
	753.2	Obstructive defects of renal pelvis & ureter	1	1
	742.4	Other specified anomalies of brain	1	1
	746.0	Anomalies of pulmonary valve	1	1
	747.1	Coarctation of aorta	0	2
	754.3	Congenital dislocation of hip	0	2
	755.2	Reduction deformities of upper limb	0	2
	275.4	Disorders of calcium metabolism	0	1
	759.2	Anomalies of other endocrine glands	0	1
	748.3	Other anomalies of larynx, trachea, & bronchus	0	1
	741.9	Spina bifida w/o hydrocephalus	0	1
	742.1	Microcephalus	0	1
	745.2	Tetralogy of Fallot	0	1
	751.2	Atresia and stenosis of large intestine, rectum, & anal canal	0	1
	754.8	Other specified nonteratogenic anomalies	0	1
	756.7	Anomalies of abdominal wall	0	1
Steuben				
	754.5	Varus deformities of feet	6	8
	752.6	Hypospadias & epispadias	5	8
	745.4	Ventricular septal defect	5	2
	753.2	Obstructive defects of renal pelvis & ureter	3	2
	755.0	Polydactyly	3	2
	758.0	Down syndrome	3	2
	747.3	Anomalies of pulmonary artery	3	1
	752.5	Undescended testicle	3	1
	742.1	Microcephalus	3	0
	750.5	Congenital hypertrophic pyloric stenosis	2	1
	753.3	Other specified anomalies of kidney	2	1
	754.3	Congenital dislocation of hip	2	1
	755.2	Reduction deformities of upper limb	2	1
	243.	Congenital hypothyroidism	2	0
	746.8	Other specified anomalies of heart	2	0
	745.5	Ostium secundum atrial septal defect	1	2
	759.8	Other specified anomalies	1	2
	757.3	Other specified anomalies of skin	0	4
	753.0	Renal agenesis & dysgenesis	0	2
	754.7	Other deformities of feet	0	2
	755.6	Other anomalies of lower limb including pelvic girdle	0	2

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County	ICD-9 Code	Description	Number in 1996	Number in 1997
Suffolk				
	745.5	Ostium secundum atrial septal defect	91	94
	752.5	Undescended testicle	90	49
	752.6	Hypospadias & epispadias	80	93
	745.4	Ventricular septal defect	77	57
	753.2	Obstructive defects of renal pelvis & ureter	60	55
	750.5	Congenital hypertrophic pyloric stenosis	50	44
	755.0	Polydactyly	43	35
	754.3	Congenital dislocation of hip	40	54
	754.5	Varus deformities of feet	36	27
	746.0	Anomalies of pulmonary valve	18	15
	746.8	Other specified anomalies of heart	16	19
	758.0	Down syndrome	15	19
Sullivan				
	752.6	Hypospadias & epispadias	6	5
	752.5	Undescended testicle	4	4
	746.8	Other specified anomalies of heart	3	0
	753.2	Obstructive defects of renal pelvis & ureter	3	0
	754.5	Varus deformities of feet	2	2
	745.4	Ventricular septal defect	2	1
	746.0	Anomalies of pulmonary valve	2	0
	750.5	Congenital hypertrophic pyloric stenosis	2	0
	755.6	Other anomalies of lower limb including pelvic girdle	2	0
	749.1	Cleft lip	2	0
	748.3	Other anomalies of larynx, trachea, & bronchus	1	3
	749.0	Cleft palate	1	2
	742.3	Congenital hydrocephalus	1	1
	744.8	Other specified anomalies of face & neck	1	1
	755.0	Polydactyly	1	1
	745.5	Ostium secundum atrial septal defect	0	2
	756.0	Anomalies of skull and face bones	0	2
	755.1	Syndactyly	0	1
	759.8	Other specified anomalies	0	1
	751.3	Hirschprung's disease & other functional disorders of colon	0	1
	333.2	Myoclonus	0	1
	743.3	Congenital cataract & lens anomalies	0	1
Tioga				
	745.5	Ostium secundum atrial septal defect	6	1
	752.5	Undescended testicle	4	3
	745.4	Ventricular septal defect	3	3
	749.2	Cleft palate with cleft lip	2	1
	524.0	Major anomalies of jaw size	2	0
	752.6	Hypospadias & epispadias	2	0
	758.0	Down syndrome	2	0
	749.1	Cleft lip	2	0
	747.3	Anomalies of pulmonary artery	1	2
	745.2	Tetralogy of Fallot	1	1
	754.3	Congenital dislocation of hip	1	1
	757.3	Other specified anomalies of skin	1	0
	749.0	Cleft palate	1	0
	743.3	Congenital cataract & lens anomalies	1	0
	742.3	Congenital hydrocephalus	1	0
	743.2	Buphthalmos	1	0
	744.0	Anomalies of ear causing impairment of hearing	1	0
	745.1	Transposition of great vessels	1	0

Section IV
Ten Most Frequently Reported Major Congenital Malformations by County, 1996-1997
Congenital Malformations Registry

County	ICD-9 Code	Description	Number in 1996	Number in 1997
Tioga (cont.)				
	746.4	Congenital insufficiency of aortic arch	1	0
	746.6	Congenital mitral insufficiency	1	0
	746.8	Other specified anomalies of heart	1	0
	747.1	Coarctation of aorta	1	0
	748.0	Choanal atresia	1	0
	751.3	Hirschprung's disease & other functional disorders of colon	1	0
	754.5	Varus deformities of feet	1	0
	756.0	Anomalies of skull and face bones	1	0
	758.3	Autosomal deletion syndromes	1	0
	759.3	Situs inversus	1	0
	753.2	Obstructive defects of renal pelvis & ureter	0	4
	751.2	Atresia and stenosis of large intestine, rectum, & anal canal	0	3
	753.1	Cystic kidney disease	0	2
	746.0	Anomalies of pulmonary valve	0	1
	750.5	Congenital hypertrophic pyloric stenosis	0	1
	753.4	Other specified anomalies of ureter	0	1
	754.6	Valgus deformities of feet	0	1
	755.6	Other anomalies of lower limb including pelvic girdle	0	1
Tompkins				
	755.6	Other anomalies of lower limb including pelvic girdle	4	3
	745.4	Ventricular septal defect	4	0
	753.2	Obstructive defects of renal pelvis & ureter	2	2
	752.6	Hypospadias & epispadias	2	1
	749.1	Cleft lip	2	1
	748.3	Other anomalies of larynx, trachea, & bronchus	2	1
	752.5	Undescended testicle	1	4
	745.5	Ostium secundum atrial septal defect	1	3
	754.5	Varus deformities of feet	1	2
	749.0	Cleft palate	1	1
	750.5	Congenital hypertrophic pyloric stenosis	1	1
	228.0	Hemangioma, any site	1	0
	754.8	Other specified nonteratogenic anomalies	1	0
	754.6	Valgus deformities of feet	1	0
	749.2	Cleft palate with cleft lip	1	0
	550.9	Inguinal hernia unspecified	1	0
	758.3	Autosomal deletion syndromes	1	0
	746.8	Other specified anomalies of heart	1	0
	747.1	Coarctation of aorta	1	0
	753.0	Renal agenesis & dysgenesis	1	0
	748.5	Agenesis, hypoplasia & dysplasia, lung	1	0
	755.5	Other anomalies of upper limb including shoulder girdle	1	0
	755.2	Reduction deformities of upper limb	1	0
	277.0	Cystic fibrosis	0	3
	746.0	Anomalies of pulmonary valve	0	2
	747.3	Anomalies of pulmonary artery	0	2
	751.4	Anomalies of intestinal fixation	0	1
	751.3	Hirschprung's disease & other functional disorders of colon	0	1
	752.8	Other specified anomalies of genital organs	0	1
	758.2	Edwards syndrome	0	1
	756.0	Anomalies of skull and face bones	0	1
	754.7	Other deformities of feet	0	1
	752.7	Indeterminate sex & pseudo-hermaphroditism	0	1
	755.0	Polydactyly	0	1
	756.5	Osteodystrophies	0	1

Section IV
Ten Most Frequently Reported Major Congenital Malformations by County, 1996-1997
Congenital Malformations Registry

County	ICD-9 Code	Description	Number in 1996	Number in 1997
Tompkins (cont.)				
	741.0	Spina bifida with hydrocephalus	0	1
	524.0	Major anomalies of jaw size	0	1
	771.2	Other congenital infections	0	1
	286.0	Congenital factor VIII disorder	0	1
	756.7	Anomalies of abdominal wall	0	1
	746.3	Congenital stenosis of aortic arch	0	1
Ulster				
	750.5	Congenital hypertrophic pyloric stenosis	8	9
	752.6	Hypospadias & epispadias	8	3
	745.4	Ventricular septal defect	6	5
	745.5	Ostium secundum atrial septal defect	6	4
	749.0	Cleft palate	6	3
	752.5	Undescended testicle	5	2
	754.3	Congenital dislocation of hip	3	8
	754.5	Varus deformities of feet	3	4
	756.0	Anomalies of skull and face bones	3	4
	742.1	Microcephalus	3	0
	755.6	Other anomalies of lower limb including pelvic girdle	2	3
	755.0	Polydactyly	0	5
Warren				
	752.6	Hypospadias & epispadias	4	2
	750.5	Congenital hypertrophic pyloric stenosis	4	1
	754.3	Congenital dislocation of hip	3	2
	745.4	Ventricular septal defect	2	3
	752.4	Anomalies of cervix, vagina & external female genitalia	2	0
	753.1	Cystic kidney disease	2	0
	753.2	Obstructive defects of renal pelvis & ureter	2	0
	749.0	Cleft palate	1	1
	753.4	Other specified anomalies of ureter	1	0
	755.2	Reduction deformities of upper limb	1	0
	754.5	Varus deformities of feet	1	0
	756.7	Anomalies of abdominal wall	1	0
	751.5	Other anomalies of intestine	1	0
	755.6	Other anomalies of lower limb including pelvic girdle	1	0
	742.2	Reduction deformities of brain	1	0
	752.0	Anomalies of ovaries	1	0
	333.2	Myoclonus	1	0
	746.9	Unspecified anomaly of heart	1	0
	743.6	Congenital anomalies of eyelids, lacrimal system and orbit	1	0
	243.	Congenital hypothyroidism	1	0
	742.1	Microcephalus	1	0
	748.0	Choanal atresia	1	0
	747.4	Anomalies of great veins	1	0
	751.0	Meckel's diverticulum	1	0
	752.5	Undescended testicle	0	3
	745.5	Ostium secundum atrial septal defect	0	2
	746.8	Other specified anomalies of heart	0	2
	758.1	Patau syndrome	0	2
	749.2	Cleft palate with cleft lip	0	2
	524.0	Major anomalies of jaw size	0	1
	753.3	Other specified anomalies of kidney	0	1
	753.0	Renal agenesis & dysgenesis	0	1
	745.0	Common truncus	0	1
	747.3	Anomalies of pulmonary artery	0	1

Section IV
Ten Most Frequently Reported Major Congenital Malformations by County, 1996-1997
Congenital Malformations Registry

County	ICD-9 Code	Description	Number in 1996	Number in 1997
Warren (cont.)				
	755.0	Polydactyly	0	1
	756.1	Anomalies of spine	0	1
	758.2	Edwards syndrome	0	1
	756.0	Anomalies of skull and face bones	0	1
	753.6	Atresia and stenosis of urethra & bladder neck	0	1
	748.5	Agenesis, hypoplasia & dysplasia, lung	0	1
Washington				
	745.4	Ventricular septal defect	5	3
	750.5	Congenital hypertrophic pyloric stenosis	5	2
	745.5	Ostium secundum atrial septal defect	4	4
	752.6	Hypospadias & epispadias	3	4
	752.5	Undescended testicle	3	1
	754.3	Congenital dislocation of hip	3	1
	524.0	Major anomalies of jaw size	2	1
	746.8	Other specified anomalies of heart	2	1
	754.5	Varus deformities of feet	2	0
	745.2	Tetralogy of Fallot	2	0
	747.0	Patent ductus arteriosus	2	0
	756.0	Anomalies of skull and face bones	1	2
	749.2	Cleft palate with cleft lip	1	1
	746.4	Congenital insufficiency of aortic valve	0	1
	756.1	Anomalies of spine	0	1
	748.3	Other anomalies of larynx, trachea, & bronchus	0	1
	243.	Congenital hypothyroidism	0	1
	749.0	Cleft palate	0	1
	753.3	Other specified anomalies of kidney	0	1
	759.8	Other specified anomalies	0	1
Wayne				
	750.5	Congenital hypertrophic pyloric stenosis	10	9
	752.6	Hypospadias & epispadias	8	4
	745.4	Ventricular septal defect	7	5
	742.3	Congenital hydrocephalus	4	0
	745.5	Ostium secundum atrial septal defect	3	2
	746.8	Other specified anomalies of heart	3	1
	752.5	Undescended testicle	2	2
	755.0	Polydactyly	2	1
	755.1	Syndactyly	2	0
	749.2	Cleft palate with cleft lip	2	0
	753.2	Obstructive defects of renal pelvis & ureter	1	4
	756.0	Anomalies of skull and face bones	1	1
	741.0	Spina bifida with hydrocephalus	1	1
	746.3	Congenital stenosis of aortic valve	1	1
	754.3	Congenital dislocation of hip	1	1
	754.6	Valgus deformities of feet	1	1
	755.2	Reduction deformities of upper limb	1	1
	756.1	Anomalies of spine	1	1
	746.0	Anomalies of pulmonary valve	0	3
	754.5	Varus deformities of feet	0	2
	758.0	Down syndrome	0	1
	756.7	Anomalies of abdominal wall	0	1
	750.3	Tracheoesophageal fistula, esophageal atresia & stenosis	0	1
	282.6	Sickle-cell anemia	0	1
	742.0	Encephalocele	0	1
	741.9	Spina bifida w/o hydrocephalus	0	1

Section IV
Ten Most Frequently Reported Major Congenital Malformations by County, 1996-1997
Congenital Malformations Registry

County	ICD-9 Code	Description	Number in 1996	Number in 1997
Wayne (cont.)				
	742.5	Other specified anomalies of spinal cord	0	1
	742.9	Unspecified anomaly, brain, spinal cord & nervous system	0	1
	743.6	Congenital anomalies of eyelids, lacrimal system and orbit	0	1
	745.1	Transposition of great vessels	0	1
	746.1	Tricuspid atresia & stenosis	0	1
	746.4	Congenital insufficiency of aortic arch	0	1
	747.1	Coarctation of aorta	0	1
	751.2	Atresia and stenosis of large intestine, rectum, & anal canal	0	1
	753.4	Other specified anomalies of ureter	0	1
	754.7	Other deformities of feet	0	1
Westchester				
	752.6	Hypospadias & epispadias	71	68
	745.4	Ventricular septal defect	57	44
	745.5	Ostium secundum atrial septal defect	45	34
	753.2	Obstructive defects of renal pelvis & ureter	35	24
	752.5	Undescended testicle	30	35
	755.0	Polydactyly	25	32
	754.3	Congenital dislocation of hip	22	15
	750.5	Congenital hypertrophic pyloric stenosis	22	14
	754.5	Varus deformities of feet	18	13
	746.0	Anomalies of pulmonary valve	14	10
Wyoming				
	745.5	Ostium secundum atrial septal defect	3	5
	745.4	Ventricular septal defect	2	3
	746.0	Anomalies of pulmonary valve	2	1
	742.4	Other specified anomalies of brain	2	0
	745.3	Common ventricle	2	0
	754.5	Varus deformities of feet	2	0
	756.0	Anomalies of skull and face bones	2	0
	752.5	Undescended testicle	1	3
	750.5	Congenital hypertrophic pyloric stenosis	1	2
	755.0	Polydactyly	1	2
	228.1	Lymphangioma, any site	1	1
	748.3	Other anomalies of larynx, trachea, & bronchus	1	1
	753.2	Obstructive defects of renal pelvis & ureter	1	1
	754.3	Congenital dislocation of hip	1	1
	755.6	Other anomalies of lower limb including pelvic girdle	1	1
	758.0	Down syndrome	1	1
	747.3	Anomalies of pulmonary artery	1	1
	749.0	Cleft palate	1	0
	742.5	Other specified anomalies of spinal cord	1	0
	277.0	Cystic fibrosis	1	0
	275.4	Disorders of calcium metabolism	1	0
	286.0	Congenital factor VIII disorder	1	0
	743.6	Congenital anomalies of eyelids, lacrimal system and orbit	1	0
	746.1	Tricuspid atresia & stenosis	1	0
	747.8	Other specified anomalies of circulatory system	1	0
	750.2	Other specified anomalies, mouth and pharynx	1	0
	751.3	Hirschprung's disease & other functional disorders of colon	1	0
	754.8	Other specified nonteratogenic anomalies	1	0
	756.5	Osteodystrophies	1	0
	758.5	Other conditions due autosomal anomalies	1	0
	758.6	Gonadal dysgenesis	1	0
	758.8	Other conditions due to sex chromosome anomalies	1	0
	746.8	Other specified anomalies of heart		

Section IV
Ten Most Frequently Reported Major Congenital Malformations by County, 1996-1997
Congenital Malformations Registry

County	ICD-9 Code	Description	Number in 1996	Number in 1997
Wyoming (cont.)				
	759.8	Other specified anomalies	0	3
	228.0	Hemangioma, any site	0	2
	746.3	Congenital stenosis of aortic arch	0	1
	748.4	Congenital cystic lung	0	1
	751.2	Atresia and stenosis of large intestine, rectum, & anal canal	0	1
	752.6	Hypospadias & epispadias	0	1
	753.0	Renal agenesis & dysgenesis	0	1
	754.6	Valgus deformities of feet	0	1
Yates				
	745.4	Ventricular septal defect	2	1
	754.3	Congenital dislocation of hip	2	1
	745.5	Ostium secundum atrial septal defect	1	1
	752.6	Hypospadias & epispadias	1	1
	749.0	Cleft palate	1	0
	343.9	Infantile cerebral palsy unspecified	1	0
	746.4	Congenital insufficiency of aortic arch	1	0
	747.3	Anomalies of pulmonary artery	1	0
	750.2	Other specified anomalies, mouth and pharynx	1	0
	754.6	Valgus deformities of feet	1	0
	757.1	Ichthyosis congenita	1	0
	752.5	Undescended testicle	0	2
	747.1	Coarctation of aorta	0	1
	750.5	Congenital hypertrophic pyloric stenosis	0	1
	742.3	Congenital hydrocephalus	0	1
	753.1	Cystic kidney disease	0	1
	753.2	Obstructive defects of renal pelvis & ureter	0	1
	753.3	Other specified anomalies of kidney	0	1
	755.4	Reduction deformities, unspecified limb	0	1

NOTES

Section V
Comparison of Selected Malformation Prevalence with Other Birth Defects Registries

Introduction to Table

The CMR relies on reports from hospitals and physicians for case ascertainment. Underreporting is an obvious concern, and the CMR monitors hospital reporting and follows up if there appears to be underreporting. In this section, CMR live birth prevalence are compared with the prevalences of two other registries, the Metropolitan Atlanta Congenital Defects Program (MACDP) [1] and the California Birth Defects Monitoring Program (CBDMP).[2] These two registries send data collection specialists to hospitals to identify and abstract records of children with malformations. To help evaluate possible underreporting, CMR prevalences of selected malformations, defined using BPA codes were compared with prevalence from these two registries. (See Appendix 4 for further information on these BPA codes.) These two programs follow children through one year of age. The CMR follows children through two years; however, more than 95% of cases are reported in the first year. Most of the malformations in this table are recognized at birth. The exceptions are fetal alcohol syndrome and some cardiac malformations.

Several malformations, including spina bifida, coarctation of the aorta, pyloric stenosis and diaphragmatic hernia have similar prevalence rates among the registries indicating that the CMR's surveillance system is effective in case ascertainment for these defects.

There are, however, apparent differences among the registries in the prevalence of some malformations. Caution should be used when comparing rates from different surveillance systems. Some differences are more likely due to differences in surveillance practices. Factors that may influence prevalence rates include case ascertainment methods, sources, and case inclusion criteria. For example, the low prevalence of anencephaly in New York State when compared to the other registries is probably due to the inclusion of stillborn infants in the MACDP and CBDMP. The CMR includes only live born children (see Section VI, Current Topics).

Variation among the registries in the rates of specific defects may also reflect demographic differences in the populations. The prevalence of Down syndrome, trisomy 18 and trisomy 13 is highly dependent upon the maternal age distribution, age-specific pregnancy rates and women's use of prenatal diagnosis and pregnancy termination. The lower live birth prevalence rates of these chromosomal abnormalities in the CMR may be partially attributable to one or more of these factors.

There are also racial and ethnic differences in the rates of specific birth defects. A report from the CBDMP for the years 1983-1990 showed that whites had the highest pyloric stenosis rate, hispanics the highest rates of neural tube defects and blacks had a higher rate of heart defects than the other races [3]. There may also be true geographic differences. A comparison of birth defect prevalences between the MACDP and CBDMP for the years 1983-1988 that adjusted for race, sex and maternal age showed regional differences in arm, hand and limb reduction defects [4].
Although there are differences in the rates of some specific defects presented in Table 5, these differences are not substantive.

Section V
Comparison of Selected Malformation Prevalence with two other Birth Defects Registries

Malformations	CMR 1996	MACDP[5] 1996	CBDMP[5] 1995
Anencephalus	0.6	2.2	2.0
Spina bifida	3.5	3.4	3.6
Hydrocephalus	7.8	8.1	5.8
Encephalocele	0.9	1.7	0.8
Microcephalus	5.8	8.8	--
An/Microphthalmos	1.4	3.4	2.8
Common truncus	0.7	1.2	0.7
Trans of great vessels	4.0	6.1	2.8
Tetralogy of Fallot	4.2	5.1	3.8
Ventricular septal defect	38.8	32.3	16.9
Hypoplastic left heart syndrome	2.0	3.2	1.8
Coarctation of aorta	4.3	4.4	4.7
Choanal atresia	2.2	2.2	1.0
Lung agenesis/hypoplasia	2.8	5.9	--
Cleft palate	6.1	6.1	7.3
Cleft lip ∀ cleft palate	8.2	9.8	10.0
Esophageal/tracheoesophageal atresia fistula	2.4	1.5	2.8
Rectal/large intestine atresia	3.3	2.2	3.4
Pyloric stenosis	17.8	14.9	17.8
Hirschsprung's disease	2.2	2.0	1.2
Biliary atresia	1.0	0.2	0.6
Renal agenesis/hypoplasia	3.0	6.1	--
Bladder exstrophy	0.3	--	0.2
Hypo/epispadias	34.1	35.0	12.5
Reduct deform of upper limb	2.7	3.9	4.1
Reduct deform of lower limb	1.4	2.0	1.3
Diaphragmatic hernia	2.4	2.7	2.5
Omphalocele	1.4	1.7	1.6
Gastroschisis	1.6	1.7	2.6
Down syndrome	8.9	12.7	12.8
Trisomy 13	0.5	1.0	0.9
Trisomy 18	0.8	2.0	2.1
Fetal alcohol syndrome	1.6	3.2	0.8
Amniotic bands	0.4	1.7	1.2

References

1. Edmonds LD, Layde PM, Levy JM, et al. Congenital malformations surveillance: two American systems. *Inter J Epidemiol* 1981; 10:247-251.

2. Grether JK. New California program monitors birth defects. *J Perinatology* 1985; 5:8-10.

3. Stierman L, Birth Defects in California: 1983-1990, California Birth Defects Monitoring Program Report Series, December, 1994.

4. Schulman J, Edmonds LD, McClern AB, et al. Surveillance for and comparison of birth defect prevelences in two geographic areas - United States 1983-1988. In: CDC Surveillance Summaries; March 19, 1993. *Morbidity and Mortality Weekly Report* 1993; 42(No. SS-1):1-7.

5. Birth defects surveillance data from selected states, A report from the National Birth Defects Prevention Network. *Teratology* 200; 61:86-159.

NOTES

Section VI
Current Topics

The National Centers for Birth Defects Research and Prevention

Birth defects are the leading cause of infant death in the United States, accounting for more than 20% of all infant deaths. They also contribute substantially to illness, long-term disability, and in some cases, an earlier death.

One of the first steps in preventing birth defects is identifying their causes; however, the causes of about 40-60% of all birth defects are still unknown. One reason is that it is difficult to conduct studies with enough cases of *specific* birth defects to identify their causes. Overcoming this difficulty requires a concerted national effort to determine what environmental, genetic, occupational, nutritional, and behavioral factors cause or contribute to birth defects.

To help reduce birth defects among U.S. babies, in 1996, Congress directed the Centers for Disease Control and Prevention (CDC) to establish the Centers of Excellence for Birth Defects Research and Prevention. This was formalized with passage of the Birth Defects Prevention Act of 1998 (Public Law 105-168). This Act authorized CDC to (1) collect, analyze, and make available data on birth defects; (2) operate regional centers that will conduct applied epidemiological research for the prevention of birth defects; and (3) provide the public with information on preventing birth defects. In1996, the New York Center was established as part of the New York State Department of Health's Congenital Malformations Registry. Currently, CDC has established centers in 7 states awarding each center $800,000 per year for 5 years. The states are Arkansas, California, Iowa, Massachusetts, New Jersey, New York and Texas.

The Centers are all participating in the National Birth Defects Prevention Study, conducting center-specific research projects, and expanding and improving birth defect surveillance systems in their states. The research done by these Centers will increase understanding, potentially dramatically, of the causes of birth defects and will provide information for developing effective birth defects prevention activities.

National Birth Defects Prevention Study

The major research effort of all of the Centers is the National Birth Defects Prevention Study (NBDPS). The NBDPS is one of the largest case-control studies of the causes of birth defects ever conducted. This study will provide information about potential causes of birth defects and a mechanism for identifying new factors or substances in the environment that are harmful to developing babies.

The study has three components. First, through existing surveillance systems, the Centers are identifying and collecting information on infants who have at least one of 30 major birth defects for which there are unknown or uncertain causes. The cases are reviewed and classified by clinical geneticists and entered into a central clinical database. The Centers also collect data on selected infants who do not have any major malformations ("controls") and compare the data to that collected for infants with birth defects ("cases"). Second, the infants' mothers are interviewed using a computer-assisted telephone interview (CATI). The interview includes questions about pregnancy and medical histories, lifestyle habits, and possible exposure of the mother or her fetus to harmful substances in the mother's workplace. The data from all sites, 16,000 interviews over a 5-year span, will be pooled electronically so that researchers can analyze the data to identify an association between risk factors and birth defects. Third, the Centers are collecting cheek cells samples from the infants and their parents. Researchers will study the DNA (genetic material) from these cheek cells to find out genetic susceptibilities to birth defects. Some of the DNA will be stored in a specimen bank at the CDC for future research as new hypotheses and improved technology emerge.

30 Major Birth Defects by Organ System Affected
Studied in the National Birth Defects Prevention Study

A birth defect is considered major if it affects survival, requires substantial medical care, or results in marked physiological or psychological impairment (handicap).

Cardiovascular: Conotruncal heart defects, Flow related heart defects, Cell death heart defects, Atrioventricular canal defects, Single ventricle, Anomalous pulmonary venous return, Ventricular septal defect - NOS, Heterotaxy
Central Nervous System: Neural tube defects, Hydrocephaly, Holoprosencephaly, Dandy-Walker malformation
Eye: Anophthalmia, Microphthalmia
Ear: Anotia, Microtia
Gastrointestinal: Esophageal atresia and Tracheoesophageal fistula, Intestinal atresia, Biliary atresia
Genitourinary: Bladder exstrophy, Renal agenesis - bilateral, Hypospadias-2nd or 3rd degree
Musculoskeletal: Diaphragmatic hernia, Gastroschisis, Omphalocele, Limb deficiency, Craniosynostosis, Cloacal exstrophy, Sacral agenesis/caudal regression
Orofacial: Choanal atresia, Orofacial clefts
Other: Amniotic band sequence

New York State Department of Health
Center for Birth Defects Research and Prevention

The New York Center's mission is to refine existing surveillance activities and to develop partnerships to conduct birth defect prevention research. The Center draws its cases for the National Birth Defects Prevention Study from two areas: (1) eight counties in western New York that are also the focus of fetal alcohol syndrome surveillance, and (2) the lower Hudson Valley Region, which covers seven counties and is also the focus of neural tube defect surveillance. Researchers from New York State Department of Health's Wadsworth Center work closely with CMR staff.

The Center's areas of special expertise include Geographic Information Systems, occupational exposure studies, environmental epidemiology, folate metabolism, mouse genetics, socioeconomic status and disease.

Key Center-Specific Projects

- Studying gene-environment interactions involved in oral clefts
- Studying the relationship between socioeconomic status and malformations
- Studying folate derivatives and other factors involved in neural tube defects
- Studying caffeine and selected birth defects
- Examining quality of care issues for children with birth defects
- Studying the relationship between congenital malformations and susceptibility to cancer

- Studying asthma medication and congenital heart malformations and other selected birth defects
- Studying maternal illnesses, such as hypertension and thyroid disease, and selected birth defects

- Studying the effects in the offspring of maternal HIV medication
- Establishing a Hemangioma Registry
- Supporting the development of Clinical Guidelines for the Work-up of Children with malformations
- Studying mutations in androgen receptor genes and hypospadias
- Active surveillance of neural tube defects and fetal alcohol syndrome
- Collaborating with other Centers on studies of pesticide exposure, occupational exposure and drinking water

Center's Partners

- Northern-Metro Chapter of March of Dimes
- Pediatric Otolaryngology Department, Children's Hospital of Buffalo
- Regional Medical Genetics Center, Westchester
- Division of Genetics, Children's Hospital of Buffalo
- Western New York Teratology Information Service
- Department of Genetics, University of Rochester

NOTES

APPENDICES

Appendix 1

Reporting Card, Congenital Malformations Registry

Congenital Malformations Registry
Confidential Case Report

New York State Department of Health
Bureau of Environmental and Occupational Epidemiology

Type or print clearly using blue or black ink.

Child's Information	**PFI Number**	**Medical Record Number**

Child's Name Last First M.I. **(DOH USE ONLY)**

AKA: If child has been identified by another name(s), enter the name(s)

Address Street City State Zip Code

Date of Birth (month/day/year)	**Birth Status**	**Birthweight** (grams)	**Sex**
__ __ / __ __ / __ __ __ __	☐ Live ☐ Still		☐ Male ☐ Female ☐ Undesignated

Race
☐ White ☐ Black or African American ☐ American Indian/Alaskan Eskimo ☐ Asian/Pacific Islander ☐ Unknown

Hispanic ☐ Yes ☐ No

Plurality
☐ Single ☐ Twin ☐ Triplet Other, specify _____

If a multiple birth, specify birth order
☐ 1st ☐ 2nd ☐ 3rd Other, specify _____

Born at this facility
☐ Yes ☐ No

If not born at this facility: Hospital of Birth City State Zip Code

Date of Discharge (month/day/year)	**Deceased**	**If deceased, date of death** (month/day/year)	**Foster/Adopted**
__ __ / __ __ / __ __ __ __	☐ Yes ☐ No	__ __ / __ __ / __ __ __ __	☐ Foster ☐ Adopted ☐ No

Diagnostic Information

	ICD Code	Narrative
1)	_____	_____
2)	_____	_____
3)	_____	_____
4)	_____	_____
5)	_____	_____
6)	_____	_____
7)	_____	_____
8)	_____	_____
9)	_____	_____
10)	_____	_____

Chromosome Studies
If yes, Karyotype _____
If pending, cytogenetic lab _____

Parents' Information

Mother's Name Last First M.I. Maiden Name

Date of Birth (month/day/year) __ __ / __ __ / __ __ __ __ **Social Security Number** __ __ __ - __ __ - __ __ __ __

Father's Name Last First M.I.

Date of Birth (month/day/year) __ __ / __ __ / __ __ __ __ **Social Security Number** __ __ __ - __ __ - __ __ __ __

Congenital Malformations Registry
Confidential Case Report

New York State Department of Health
Bureau of Environmental and Occupational Epidemiology

Type or print clearly using blue or black ink.

Reporting Source			
(Stamp Acceptable) Name Street Address City State Zip Code			**Check here if you need more:** ☐ Forms ☐ Envelopes
CMR Registrar	Last	First	Phone
Attending Physician	Last	First	Phone
Pediatrician	Last	First	Phone
Patient transferred from another facility:	(Enter name of facility)		
Patient transferred to another facility:	(Enter name of facility)		

Mail completed form in sealed envelope to:

New York State Department of Health
Bureau of Environmental and Occupational Epidemiology
Congenital Malformations Registry
Flanigan Square, Room 200
547 River Street
Troy, NY 12180-2216
Telephone: (518) 402-7990

Appendix 2

Classification of Codes

Congenital malformations have traditionally been divided into categories of "major" and "minor". A major anomaly has an adverse effect on the individual's health, functioning or social acceptability. A minor anomaly is generally considered of limited social or medical significance. While minor anomalies in themselves do not greatly affect the child, they can be related to major anomalies or be indications of certain syndromes.[1,2]

The division between major and minor is far from perfect. No standard lists or definitions exist. We used several sources, including the practices of other registries, to develop a list of minor anomalies.[3, 4, 5] One serious problem in making this distinction is that some ICD-9-CM codes include major and minor malformations under the same code. A more specific coding scheme that eliminates most of these problems has been adopted.

Following is a general listing of conditions included in this report and their classification. A few codes are not listed since they contain only a very few cases. Reporting hospitals receive a CMR Handbook with a complete, detailed list of reportable anomalies.

Major Malformations

090.0 – 090.9	Congenital Syphilis
658.8	Amniotic Bands
740 - 759*	Congenital Anomalies
760.71	Fetal Alcohol Syndrome
771.0 - 771.2	Congenital Infections: including rubella, cytomegalovirus toxoplasmosis and herpes simplex

*See list of minor and excluded codes

Minor Malformations

214	Lipoma
216	Benign neoplasm of skin
228.01	Hemangioma of skin
550	Inguinal hernia in males
553.1	Umbilical hernia
743.65	Specified congenital anomalies of lacrimal passages
744.1	Accessory auricle
744.29	Other specified anomalies of ear
744.3	Unspecified anomaly of ear
744.4	Branchial cleft cyst
744.89	Other specified anomalies of face and neck
744.9	Other unspecified anomalies of face and neck
747.0	Patent ductus arteriosis, if birth weight <1500 grams
747.5	Single umbilical artery
752.41	Embryonic cyst of cervix, vagina and external female genitalia
752.42	Imperforate hymen
752.5	Undescended testicle, if birth weight < 2500 grams
754.61	Congenital pes planus
755.0	Polydactyly
755.11, 755.13	Syndactyly without fusion of bone
757.2	Dermatoglyphic anomalies
757.32	Vascular hamartomas
757.33	Congenital pigmentation anomalies of skin
757.39	Other anomalies of skin
757.4	Specified anomalies of hair
757.5	Specified anomalies of nails
757.6	Specified anomalies of breast
757.8	Other specified anomalies of integument
757.9	Unspecified anomalies of the integument

Exclusions

750.0	Tongue tie
758.4	Balanced autosomal translocation in normal individual
778.6	Congenital hydrocele

References

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3. Merlob P, Papier CM, Klingberg MA, Reisner SH. Incidence of congenital malformations in the newborn, particularly minor abnormalities. In: Marois, ed. *Prevention of physical and mental congenital defects, Part C: Basic and medical sciences, education and future strategies. Proceedings of a conference of the Institut de la Vie***.** New York: Alan R. Liss, 1985:51-53.

4. Myrianthopoulos NC, Chung CS. Congenital malformations in singletons: epidemiologic survey. Birth Defects: *Original Article Series*, 1974; X: 2-3, 51-58.

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Appendix 3

Birth Certificate Matching

Birth certificate matching is a vital part of registry activities. This serves to verify the individual's identity and distinguish him or her from all others and provides additional information about the baby and the mother. The matching is used to determine maternal residence at birth and to verify race and birthweight. Matched cases provide a basis to calculate population-based rates. It is critical to match a high percentage of cases to calculate rates accurately and to conduct meaningful surveillance.

Birth certificate matching is carried out by a computer program that compares the birth certificate tape for a given year to the CMR file of cases who were born in that year. The files are compared on several variables until (1) a match is found, (2) a possible match is found or (3) the list is exhausted without finding a match.

Possible matches are reviewed by CMR staff and a decision made about whether there is a match. Unmatched cases are checked further to see if data items have been correctly keyed and all possible aliases have been identified. An online search of the birth certificate files is done and certificates on file at the Vital Records office are reviewed to find unmatched cases. However, review of actual certificates is possible only for children born outside New York City since New York City birth certificates are not on file in Albany. New York City maintains its own vital records files.

The matching process is repeated until about 95% of reported cases are matched. This is a compromise between completeness and efficiency. After about 90% of cases are matched, each additional percentage requires greater and greater effort. The ability to review a copy of the birth certificate greatly enhances the chance of making a match. Matching is more complete for cases born in the state outside New York City than for New York City cases.

Appendix 4

BPA Codes

Many birth defects registries use a coding system modified from the British Pediatric Association (BPA). This coding system provides more specificity than the ICD-9 system. The Centers for Disease Control and Prevention Metropolitan Atlanta Congenital Defects Program (MACDP) has developed codes that group conditions. The table below shows the MACDP codes and the corresponding BPA and ICD-9 codes. The ICD-9 code may include conditions others than those specified by the BPA code. For example, ICD-9 code 756.7 includes both gastroschisis and omphalocele, but the BPA code allows these conditions to be distinguished.

MACDP Code	Condition	ICD-9	BPA 5-Digit Code
CENTRAL NERVOUS SYSTEM			
A01	Anencephaly	740.0, 740.1, 740.2	740.00, 740.02, 740.03, 740.10, 740.20, 740.21, 740.29
A02	Spina Bifida with Hydrocephaly	741.0	741.00, 741.01, 741.02, 741.03, 741.04, 741.05, 741.06, 741.07, 741.08, 741.09
A03	Spina Bifida without Hydrocephaly	741.9	741.90, 741.91, 741.92, 741.93, 741.94, 741.98, 741.99, 742.00, 742.08, 742.09
A13	Encephalocele	742.0	742.00, 742.08, 742.09
A15	Hydrocephaly	742.3	742.30, 742.31, 742.38, 742.39
A16	Microcephalus	742.1	742.10
EYE / EAR			
B01	Anophthalmia, Microphthalmia	743.0, 743.1	743.00, 743.10
B03	Glaucoma	743.2	743.20, 743.21, 743.22
B04	Cataract		743.32
B54	Ear anomaly with hearing loss	744.0	744.00, 744.01, 744.02, 744.03, 744.09, 744.21
CARDIAC			
D01	Truncus arteriosus	745.0	745.00, 745.01
D02	Transposition of great vessels	745.1	745.10, 745.11, 745.12, 745.18, 745.19
D03	Tetralogy of Fallot	745.2	745.20, 745.21, 746.84
D04	Single ventricle	745.3	745.30
D05	VSD	745.4	745.40, 745.41, 745.48, 745.49
D52	Hypoplastic left heart	746.7	746.70
D53	Total anomalous pulmonary venous return	747.41	747.42
RESPIRATORY			
E01	Choanal atresia	748.0	748.00
E06	Agenesis of lung	748.5	748.50, 748.51
CLEFTS			
F01	Cleft palate	749.0	749.00, 749.01, 749.02, 749.03, 749.04, 749.05, 749.06, 749.07, 749.09
F02	Cleft lip with or without cleft palate	749.0, 749.2, 750.5	749.10, 749.11, 749.12, 749.19, 749.20, 749.21, 749.22, 749.29, 749.51

MACDP Code	Condition	ICD-9	BPA 5-Digit Code
GASTRO-INTESTINAL			
F14	Stenosis or atresia of duodenum	751.1	751.10
F15	Other stenosis or atresia of small intestine	751.1	751.11, 751.12, 751.19
F16	Stenosis or atresia of rectum or anus	751.2	751.21, 751.22, 751.23, 751.24
F17	Hirschsprung□s Disease	751.3	751.30, 751.31, 751.32, 751.33
F18	Malrotation of intestine	751.4	751.40, 751.41, 751.42, 751.49
F21	Biliary atresia	751.61	751.65
GENITO-URINARY			
H01	Renal agenesis	753.0	753.00, 753.01
H06	Obstruction of kidney or ureter	753.3	753.20, 753.21, 753.22, 753.29, 753.40, 753.42
H09	Bladder or urethra obstruction	753.6	753.60, 753.61, 753.62, 753.63
MUSCULOSKELETAL			
J02	Curvature of spine (scoliosis or lordosis)	754.2	754.20, 754.21, 754.22
J03	Dislocation of hip	754.3	754.30
J11	Arthrogryposis multiplex congenita	754.89	755.80
K01	Reduction deformity - upper limb	755.2	755.20, 755.21, 755.22, 755.23, 755.24, 755.25, 755.26, 755.27, 755.28, 755.29
K02	Reduction deformity - lower limb		755.30, 755.31, 755.32, 755.33, 755.34, 755.35, 755.36, 755.37, 755.38, 755.39
K05	Amniotic bands	658.8	658.80
N01	Diaphragmatic hernia	756.6	756.61
N02	Omphalocele	756.7	756.70
N04	Gastroschisis	756.7	756.71
SYNDROMES			
R01	Down Syndrome	758.0	758.00, 758.01, 758.02, 758.03, 758.04, 758.09
R02	Patau Syndrome (Trisomy 13)	758.1	758.10, 758.11, 758.12, 758.13, 758.19
R03	Edwards Syndrome (Trisomy 18)	758.2	758.20, 758.21, 758.23, 758.29
S02	Fetal Alcohol Syndrome	760.71	760.71
W03	Conjoined twins	759.4	759.40, 759.41, 759.42, 759.43, 759.44, 759.48, 759.49

Appendix 5

Glossary of Terms*

Agenesis Absence of part(s) of the body.

Agenesis, aplasia, or hypoplasia of the lung The absence or incomplete development of a lung or lung tissue.

Anencephaly Congenital absence of the skull, with cerebral hemispheres completely missing or reduced to small masses attached to the base of the skull. Anencephaly is not compatible with life.

Aniridia The complete absence of the iris of the eye or a defect of the iris. Can be congenital or traumatically induced.

Anopthalmia A developmental defect characterized by complete absence of the eyes, or by the presence of vestigial eyes.

Anotia A congenital absence of one or both ears.

Aortic valve stenosis A cardiac anomaly characterized by a narrowing or stricture of the aortic valve. This condition causes abnormal cardiac circulation and pressure in the heart during contractions. This condition can be repaired surgically in some cases.

Atresia Imperforation; absence or closure of a normal opening.

Atrial Septal Defect A congenital cardiac malformation in which there are one or several openings in the atrial septum (muscular and fibrous wall between the right and left atria) allowing a mixing of oxygenated and unoxygenated blood. The openings vary in size and may resolve without treatment or may require surgical treatment. Also called *ostium secundum defect.*

Biliary atresia A congenital absence or underdevelopment of one or more of the ducts in the biliary tract. Correctable surgically.

Bladder extrophy Incomplete closure of the anterior wall of the bladder and the abdominal cavity. The upper urinary tract is generally normal. Often associated with anorectal and genital malformations, and epispadias. Affected persons are at a markedly increased risk of bladder carcinoma (squamous cell). This condition is usually corrected surgically after birth.

Cataract An opacity (clouding) of the lens of the eye.

Choanal atresia or stenosis A congenital anomaly in which a bony or membranous formation blocks the passageway between the nose and the pharynx. This defect is usually repaired surgically after birth. Bilateral Choanal atresia is a surgical emergency.

Cleft lip The congenital failure of the fetal components of the lip to fuse or join, forming a groove or fissure in the lip. Infants with this condition can have difficulty feeding, and may use assistive devices for feeding. This condition is corrected when the infant can tolerate surgery.

Cleft palate The congenital failure of the palate to fuse properly, forming a grooved depression or fissure in the roof of the mouth. This defect varies in degree of severity. The fissure can extend into the hard and soft palate and into the nasal cavities. Infants with this condition have difficulty feeding, and may use assistive devices for feeding. Surgical correction is begun as soon as possible. Children with cleft palates are at high risk for hearing problems due to ear infections.

Coarctation of the aorta Localized narrowing of the aorta. This condition causes abnormal cardiac circulation and pressure in the heart during contractions. This condition can vary from mild to severe. Surgical correction is recommended even for mild defects.

Common Truncus Ateriosus A congenital heart defect in which the common arterial trunk fails to divide into pulmonary artery and aorta. This is corrected surgically.

Confidence interval (95%) The interval that contains the true prevalence (which we can only estimate) 95% of the time.

Congenital Existing at or dating from birth.

Congenital hip dislocation A congenital defect in which the head of the femur does not articulate with the acetabulum of the pelvis because of an abnormal shallowness of the acetabulum. Treatment in early infancy consists of bracing of the joint to cause a deepening of the acetabulum.

Conjoined Twins Monozygotic twins who are physically united at birth. The defect can range from a superficial connection to one in which only a single body part is duplicated. Classified as symmetrical or asymmetrical by the degree of separation and development.

Craniosynostosis A premature ossification (closing) of the cranial sutures before birth or soon after birth. This condition is occasionally associated with other skeletal defects. If no surgical correction is made, the growth of the skull is inhibited, and the head is deformed. The eyes and the brain are often damaged.

Diaphragmatic hernia A failure of the diaphragm to form completely, leaving a hole. Abdominal organs can protrude through the hole into the chest cavity and interfere with development of the heart and lungs. Usually life-threatening and requires emergent surgery.

Down syndrome (Trisomy 21) The chromosomal abnormality characterized by an extra copy of chromosome 21. In rare cases this syndrome is caused by *translocation.* The extra copy can be free-lying, or can be attached to some other chromosome, most frequently number 14. Down syndrome can occur in *mosaic.* So that there is a population of normal cells and a population of trisomy 21 cells. Down syndrome is characterized by moderate to severe mental retardation, sloping forehead, small ear canals, flat bridged nose and short fingers and toes. One third of infants have congenital heart disease, and one third have duodenal atresia. (Both can be present in the same infant.) Affected people can survive to middle or old age. There is an increased incidence of Alzheimer disease in adults with Down syndrome.

Ebstein anomaly A congenital heart defect in which the tricuspid valve is displaced downward into the right ventricle causing abnormal patterns of cardiac circulation.

Edwards syndrome (Trisomy 18) The chromosomal abnormality characterized by an extra copy of chromosome 18. The extra chromosome can be free lying or attached to another chromosome. Trisomy 18 can occur in mosaic. Edwards syndrome is characterized by mental retardation, neonatal hepatitis, low-set ears, skull malformation and short digits. Cardiac and renal anomalies are also common.

Survival for more than a few months is rare.

Encephalocele The protrusion of the brain substance through a defect in the skull.

Endocardial cushion defect A variety of septal defects (malformations of the walls separating the two atria and two ventricles of the heart) resulting from imperfect fusion of the endocardial cushions in the embryonic heart.

Epispadias A congenital defect in which the urinary meatus (urinary outlet) opens above (dorsal to) the normal position. The urinary sphincters are defective, so incontinence does occur. Surgical correction is aimed at correcting incontinence and permitting sexual functioning. The corresponding defect in females is rare. See also Hypospadias.

Esophageal Stenosis or Atresia A narrowing or incomplete formation of the esophagus. Usually a surgical emergency. Frequently associated with a Tracheoesophageal Fistula.

Fetal alcohol syndrome A constellation of physical abnormalities (including characteristic abnormal facial features and growth retardation), and problems of behavior and cognition in children born to mothers who drank alcohol during pregnancy.

Fistula An abnormal passage from an internal organ to the body surface or between two internal organs or structures.

Gastroschisis A congenital opening of the abdominal wall with protrusion of the intestines. This condition is surgically treated. Contrast with Omphalocele, below.

Hirschsprung disease The congenital absence of autonomic ganglia (nerves controlling involuntary and reflexive movement) in the muscles of the colon. This results in immobility of the intestines and may cause obstruction or stretching of the intestines. This condition is repaired surgically in early childhood by the removal of the affected portion of the intestine.

Holoprosencephaly Failure of the brain to develop into two equal halves, so there is structural abnormality of the brain. There may be associated midline facial defects including cyclopia (fusion of the eye orbits into a single cavity containing one eye) in severe cases. About half the cases are probably due to a single gene defect (the HPE gene). Frequently occurs with Trisomy 13.

Hydrocephalus The abnormal accumulation of fluid within the spaces of the brain.

Hyperplasia Overgrowth characterized by an increase in the number of cells of a tissue.

Hypoplasia A condition of arrested development in which an organ or part remains below the normal size or in an immature state.

Hypoplastic left heart syndrome Atresia, or marked hypoplasia, of the aortic opening or valve, with hypoplasia of the ascending aorta and defective development of the left ventricle (with mitral valve atresia). This condition can be surgically repaired in a series of three procedures over a period of one year. Transplantation is also a treatment. This condition is usually fatal in the first month of life if not treated.

Hypospadias A congenital defect in which the urinary meatus (urinary outlet) is on the underside of the penis or on the perineum (area between the genitals and the anus). The urinary sphincters are not defective so incontinence does not occur. The condition may be surgically corrected if needed for cosmetic, urologic, or reproductive reasons. The corresponding defect in women is rare. *See also epispadias.*

Limb defects See Reduction deformities.

Meninges Membranes that cover the brain and spinal cord.

Microcephaly The congenital smallness of the head, with corresponding smallness of the brain.

Microphthalmia The congenital abnormal smallness of one or both eyes. Can occur in the presence of other ocular defects.

Microtia A small or maldeveloped external ear and atretic or stenotic external auditory canal.

Mosaic In genetics, this refers to an individual organism that has two or more kinds of genetically different cell types. The degree of abnormality depends on the type of tissue containing affected cells. Individuals may vary from near normal to full manifestation of the genetic syndrome. Can occur in any chromosome abnormality syndrome.

Neural tube defect A defect resulting from failure of the neural tube to close in the first month of pregnancy. The major conditions include anencephaly, spina bifida, and encephalocele.

Obstructive Genitourinary Defect Stenosis or atresia of the urinary tract at any level. Severity of the defect depends largely upon the level of the obstruction. Urine accumulates behind the obstruction and damages the organs.

Omphalocele The protrusion of an organ into the umbilicus. The defect is usually closed surgically soon after birth. Contrast with Gastroschisis.

Patau Syndrome (Trisomy 13) The chromosomal abnormality caused by a extra chromosome 13. The extra copy can be free-lying, or can be attached to some other chromosome. Patau syndrome can occur in *mosaic* so that there is a population of normal cells and a population of trisomy 13 cells. Patau syndrome is characterized by impaired midline facial development, cleft lip and palate, polydactyly and mental retardation. Most infants do not survive beyond 6 months of life.

Patent ductus arteriosus A blood vessel between the pulmonary artery and the aorta. This is normal in fetal life, but can cause problems after birth, particularly in premature infants. This condition causes abnormal cardiac circulation and pressure in the heart during contractions. The vast majority close spontaneously and cause no problems. Medical or surgical correction may be done. This is only an abnormality if it causes significant medical problems.

Pulmonary artery anomaly Abnormality in the formation of the pulmonary artery such as stenosis or atresia. See also common truncus.

Pulmonary valve atresia or stenosis A congenital heart condition characterized by absence or constriction of the pulmonary valve. This condition causes abnormal cardiac circulation and pressure in the heart during contractions. This condition can vary from mild to severe. Mild forms are relatively well tolerated and require no intervention. More severe forms are surgically corrected.

Pyloric stenosis A narrowing of the pyloric sphincter at the outlet of the stomach. This causes a blockage of food from the stomach into the small intestine. Usually treated surgically.

Reduction defects of the lower limbs The congenital absence of a portion of the lower limb. There are two general types of defect, transverse and longitudinal. Transverse defects appear like amputations, or like missing segments of the limb. Longitudinal defects are missing rays of the limb (for example, a missing tibia and great toe).

Reduction defects of the upper limbs The congenital absence of a portion of the upper limb. There are two general types of defect, transverse and longitudinal. Transverse defects appear like amputations, or like missing segments of the limb. Longitudinal defects are missing rays of the limb (for example, a missing radius and thumb).

Renal agenesis or dysgenesis The failure, or deviation, of embryonic development of the kidney.

Spina bifida A neural tube defect resulting from failure of the spinal neural tube to close. The spinal cord and/or meninges may or may not protrude. This usually results in damage to the spinal cord with paralysis of the involved limbs. Includes myelomeningocele (involving both spinal cord and meninges) and meningocele (involving just the meninges).

Stenosis A narrowing or constriction of the diameter of a bodily passage or orifice.

Stenosis or atresia of large intestine, rectum and anus The absence, closure or constriction of the large intestine, rectum or anus. Can be surgically corrected or bypassed.

Stenosis or atresia of the small intestine A narrowing or incomplete formation of the small intestine obstructing movement of food through the digestive tract.

Tetralogy of Fallot A congenital cardiac anomaly consisting of four defects: ventricular septal defect, pulmonary valve stenosis or atresia, displacement of the aorta to the right, and hypertrophy of right ventricle. The condition is corrected surgically.

Tracheoesophageal fistula An abnormal passage between the esophagus and trachea. Leads to pneumonia. Corrected surgically. It is frequently associated with esophageal atresia.

Translocation The rearrangement of genetic material within the same chromosome or the transfer of a segment of one chromosome to another one. People with balanced translocations do not always manifest genetic syndromes, but may be carriers of genetic syndromes and can have children with unbalanced translocations. Can occur with any chromosomal anomaly syndrome.

Transposition of the great vessels A congenital malformation in which the aorta arises from the right ventricle and the pulmonary artery from the left ventricle (opposite of normal), so that the venous return from the peripheral circulation is recirculated without being oxygenated in the lungs. Immediate surgical correction is needed. When this is not associated with other cardiac defects, and not corrected, it is fatal.

Tricuspid valve atresia or stenosis A congenital cardiac condition characterized by the absence or constriction of the tricuspid valve. The opening between the right atrium and right ventricle is absent or restricted, and normal circulation is not possible. This condition is often associated with other cardiac defects. This condition is surgically corrected depending on the severity.

Trisomy A chromosomal abnormality characterized by one more than the normal number of chromosomes. Normally, cells contain two of each chromosome. In trisomy, cells contain three copies of a specific chromosome.

Trisomy 13 See Patau Syndrome.

Trisomy 18 See Edwards Syndrome.

Trisomy 21 See Down Syndrome.

Truncus Arteriosus See Common Truncus.

Ventricular Septal Defect (VSD) A congenital cardiac malformation in which there are one or several openings in the ventricular septum (muscular and fibrous wall between the right and left ventricle *or right and left lower chambers of the heart*) allowing a mixing of oxygenated and unoxygenated blood. The openings vary in size and may resolve without treatment or require surgical treatment.

*Courtesy of the Texas Birth Defects Monitoring Division February 1999

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