New York State Department of Health

Congenital Malformations Registry

Annual Report

Statistical Summary of Children Born in 1994 and Diagnosed Through 1996

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Year of Publication 1998

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Summary

This report presents rates of congenital malformations occurring among the 277,945 children who were born alive to New York State residents in 1994. The children reported with a major congenital malformation represent 3.4% of live births. Males had a higher rate of major congenital malformations than females (3.9% versus 2.9%), and black children had a higher major malformation rate than white children (3.9% versus 3.3%).

Section I of this report presents demographic characteristics of children reported to the registry, number of malformations and age at diagnosis. 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.

This is the twelfth cohort report from the Congenital Malformations Registry. Reports are also available for the 1983-1993 birth cohorts. The statistics in this report are not comparable to reports before 1992. Cohort reports for the years 1989 to 1991 were not based on birth certificate matched cases. For this report, birth certificates were used to supplement or correct reported data. Birth certificate matching also helped eliminate duplicate cases reported under different names and nonresident births. In addition, 1992 was the first year that the registry used a new coding system, which allows for more specificity. For previous years, ICD-9 codes were used.

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 congenital 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 revealed 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. The CMR relies upon physicians to verify diagnoses initially suspected in the hospital but confirmed on an outpatient basis, and to clarify nonspecific diagnoses 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 (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.

1994 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 better 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. Some studies have found rates of minor congenital anomalies as high as 21%[8].

The statistics in this report are **not** comparable to reports prior to 1992. The 1994 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. 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 1994 and diagnosed before their second birthday. Reports are also available for the 1984 through 1993 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 1994 population of New York State was about 18.3 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 1994, there were 277,945 resident live births reported to the state's vital registration, 21% to black mothers, and 19.2% to Hispanic mothers. The race of the child is based on race of the mother only. Nearly 47% of live births were to New York City residents.

References

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8. 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.

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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 1994 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.

About 80% of children reported with major malformations have only one major malformation (Table 1). The overall occurrences of major malformations are 3.4% of live births. Male children have a higher rate of major malformations than female children (3.9% versus 2.9%, Table 2). This difference is consistent within different racial groups. The rates for major malformations are somewhat higher for black than for white children (3.9% versus 3.3%). The major malformation rate among children with residence at birth in New York State excluding New York City was similar to children with residence in New York City (3.3% versus 3.5%). The smaller number of births in the "other" racial category makes these rates difficult to interpret.

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 (Table 3). All race-sex groups for children with multiple major malformations showed little variation (Table 4).

Most major malformations were diagnosed within the first three days of life for all races (75.7%, Table 5). The percentage diagnosed within the first three days was slightly higher for white (76.7%) than for black children (73.9%). The proportion of diagnoses made at age six months or later is somewhat greater for black children (6.3%) than for white children (5.6% Table 5).

Section I - Table 1

1994 Births - New York State Residents

Number of Major Malformations Per Child

Number of Malformations	Number of Children	Percent
All Children	9,455	100.0**
1	7,396	78.2
2	1,220	12.9
3	426	4.5
4	180	1.9
5	97	1.0
6	52	0.5
7	26	0.3
8	26	0.3
9	12	0.1
10	9	0.1
11	3	*
12	5	0.1
13	2	*
14	1	*

* Less than 0.05%

** Total may not add to 100 due to rounding

Section 1 - Table 2
1994 Births - New York State Residents
Percent of Live Births With One Or More Major Malformations
Sex by Race and Residence

Race and Residence	Both Sexes			Males			Females		
	Infants	Total Births	%	Infants	Total Births	%	Infants	Total Births	%
New York State									
All Races‡	9,455	277,945	3.4	5,566	141,917	3.9	3,885	136,021	2.9
White	6,676	202,071	3.3	4,001	103,434	3.9	2,672	98,631	2.7
Black	2,268	57,880	3.9	1,270	29,229	4.3	998	28,651	3.5
Other	425	15,528	2.7	245	7,983	3.1	179	7,544	2.4
NYS Excluding NYC									
All Races‡	4,967	148,618	3.3	3,042	75,889	4.0	1,921	72,722	2.6
White	4,212	128,012	3.3	2,606	65,522	4.0	1,603	62,484	2.6
Black	621	15,587	4.0	361	7,843	4.6	260	7,744	3.4
Other	102	4,081	2.5	60	2,044	2.9	41	2,036	2.0
New York City									
All Races‡	4,488	129,327	3.5	2,524	66,028	3.8	1,964	63,299	3.1
White	2,464	74,059	3.3	1,395	37,912	3.7	1,069	36,147	3.0
Black	1,647	42,293	3.9	909	21,386	4.3	738	20,907	3.5
Other	323	11,447	2.8	185	5,939	3.1	138	5,508	2.5

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

Section 1 - Table 3
1994 Births - New York State Residents
Percent of Live Births With One Major Malformation
Sex by Race and Residence

Race and Residence	Both Sexes			Males			Females		
	Infants	Total Births	%	Infants	Total Births	%	Infants	Total Births	%
New York State									
All Races‡	7,396	277,945	2.7	4,454	141,917	3.1	2,942	136,021	2.2
White	5,229	202,071	2.6	3,218	103,434	3.1	2,011	98,631	2.0
Black	1,784	57,880	3.1	1,015	29,229	3.5	769	28,651	2.7
Other	319	15,528	2.1	182	7,983	2.3	137	7,544	1.8
NYS Excluding NYC									
All Races‡	3,872	148,618	2.6	2,436	75,889	3.2	1,436	72,722	2.0
White	3,280	128,012	2.6	2,089	65,522	3.2	1,191	62,484	1.9
Black	490	15,587	3.1	289	7,843	3.7	201	7,744	2.6
Other	74	4,081	1.8	44	2,044	2.2	30	2,036	1.5
New York City									
All Races‡	3,524	129,327	2.7	2,018	66,028	3.1	1,506	63,299	2.4
White	1,949	74,059	2.6	1,129	37,912	3.0	820	36,147	2.3
Black	1,294	42,293	3.1	726	21,386	3.4	568	20,907	2.7
Other	245	11,447	2.1	138	5,939	2.3	107	5,508	1.9

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

Section 1 - Table 4
1994 Births - New York State Residents
Percent of Live Births With Two Or More Major Malformations
Sex by Race and Residence

Race and Residence	Both Sexes			Males			Females		
	Infants	Total Births	%	Infants	Total Births	%	Infants	Total Births	%
New York State									
All Races‡	2,059	277,945	0.7	1,112	141,917	0.8	943	136,021	0.7
White	1,447	202,071	0.7	783	103,434	0.8	661	98,631	0.7
Black	484	57,880	0.8	255	29,229	0.9	229	28,651	0.8
Other	106	15,528	0.7	63	7,983	0.8	42	7,544	0.6
NYS Excluding NYC									
All Races‡	1,095	148,618	0.7	606	75,889	0.8	485	72,722	0.7
White	932	128,012	0.7	517	65,522	0.8	412	62,484	0.7
Black	131	15,587	0.8	72	7,843	0.9	59	7,744	0.8
Other	28	4,081	0.7	16	2,044	0.8	11	2,036	0.5
New York City									
All Races‡	964	129,327	0.7	506	66,028	0.8	458	63,299	0.7
White	515	74,059	0.7	266	37,912	0.7	249	36,147	0.7
Black	353	42,293	0.8	183	21,386	0.9	170	20,907	0.8
Other	78	11,447	0.7	47	5,939	0.8	31	5,508	0.6

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

Section 1 - Table 5
1994 Births - New York State Residents
Age at Earliest Diagnosis in Children With Major Malformations
Race and Residence

Age at Diagnosis and Residence	All Races		White		Black		Other	
	Number	%	Number	%	Number	%	Number	%
New York State								
All Ages‡	9,455	100.0	6,676	100.0	2,268	100.0	425	100.0
Less than 3 days	7,159	75.7	5,118	76.7	1,676	73.9	299	70.3
3 to 29 days	961	10.2	664	9.9	238	10.5	51	12.0
30 to 182 days	793	8.4	522	7.8	211	9.3	53	12.5
6 months to 2 yrs.	542	5.7	372	5.6	143	6.3	22	5.2
NYS Excluding NYC								
All Ages‡	4,967	100.0	4,212	100.0	621	100.0	102	100.0
Less than 3 days	3,825	77.0	3,270	77.6	453	72.9	74	72.5
3 to 29 days	440	8.9	368	8.7	62	10.0	7	6.9
30 to 182 days	425	8.6	343	8.1	65	10.5	16	15.7
6 months to 2 yrs.	277	5.6	231	5.5	41	6.6	5	4.9
New York City								
All Ages‡	4,488	100.0	2,464	100.0	1,647	100.0	323	100.0
Less than 3 days	3,334	74.3	1,848	75.0	1,223	74.3	225	69.7
3 to 29 days	521	11.6	296	12.0	176	10.7	44	13.6
30 to 182 days	368	8.2	179	7.3	146	8.9	37	11.5
6 months to 2 yrs.	265	5.9	141	5.7	102	6.2	17	5.3

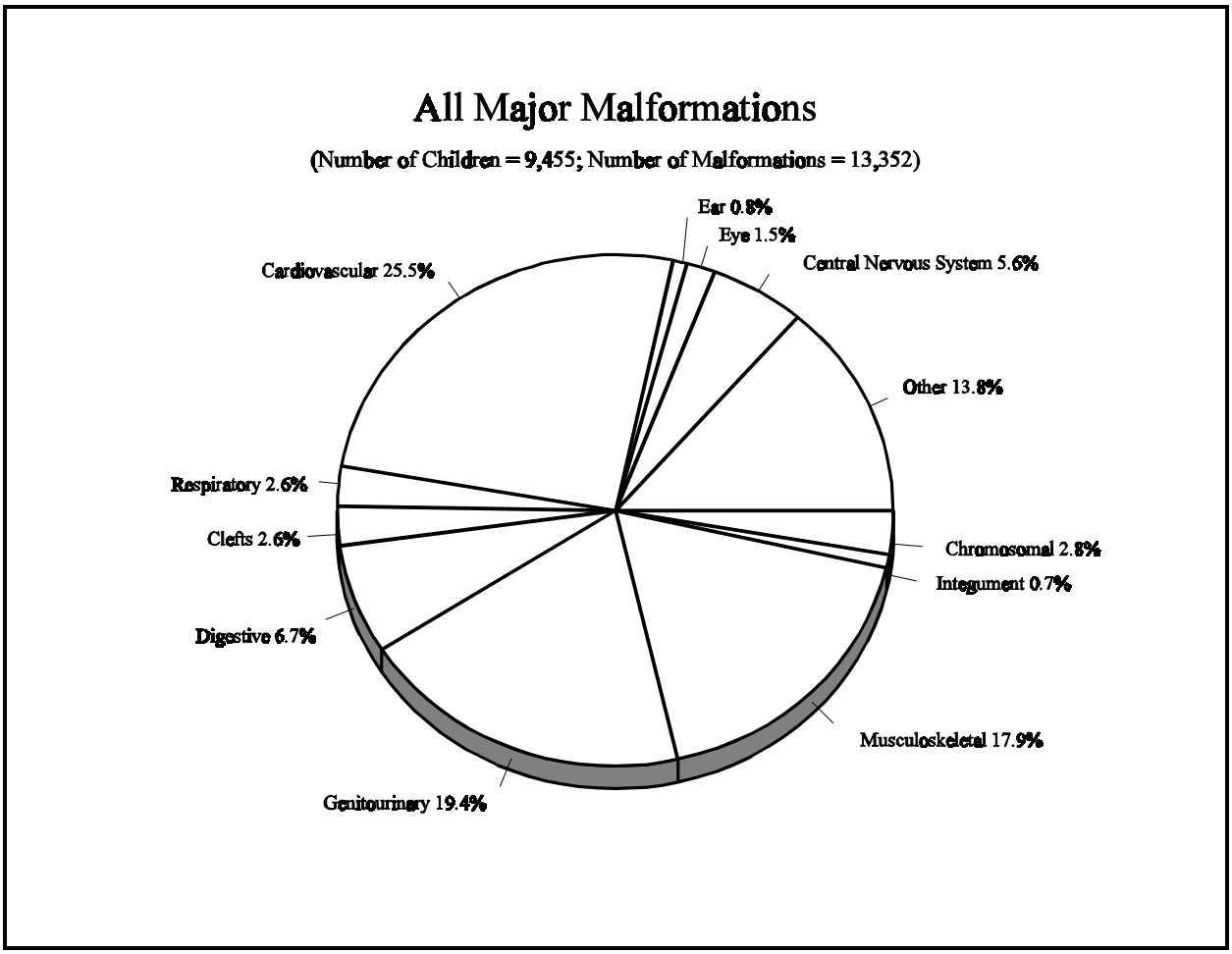
‡Total includes unknowns within each category, thus row and column figures may not sum to totals.

Section II
Major Congenital Malformations by Organ System

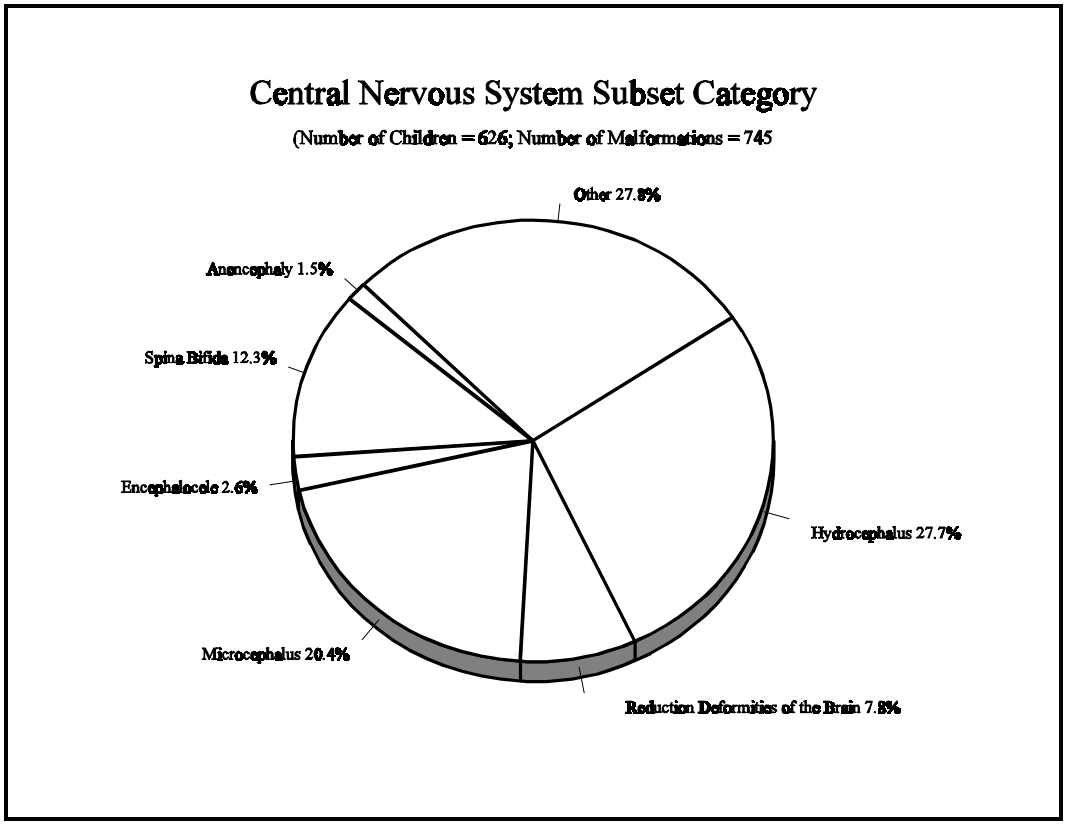
Introduction to Figures

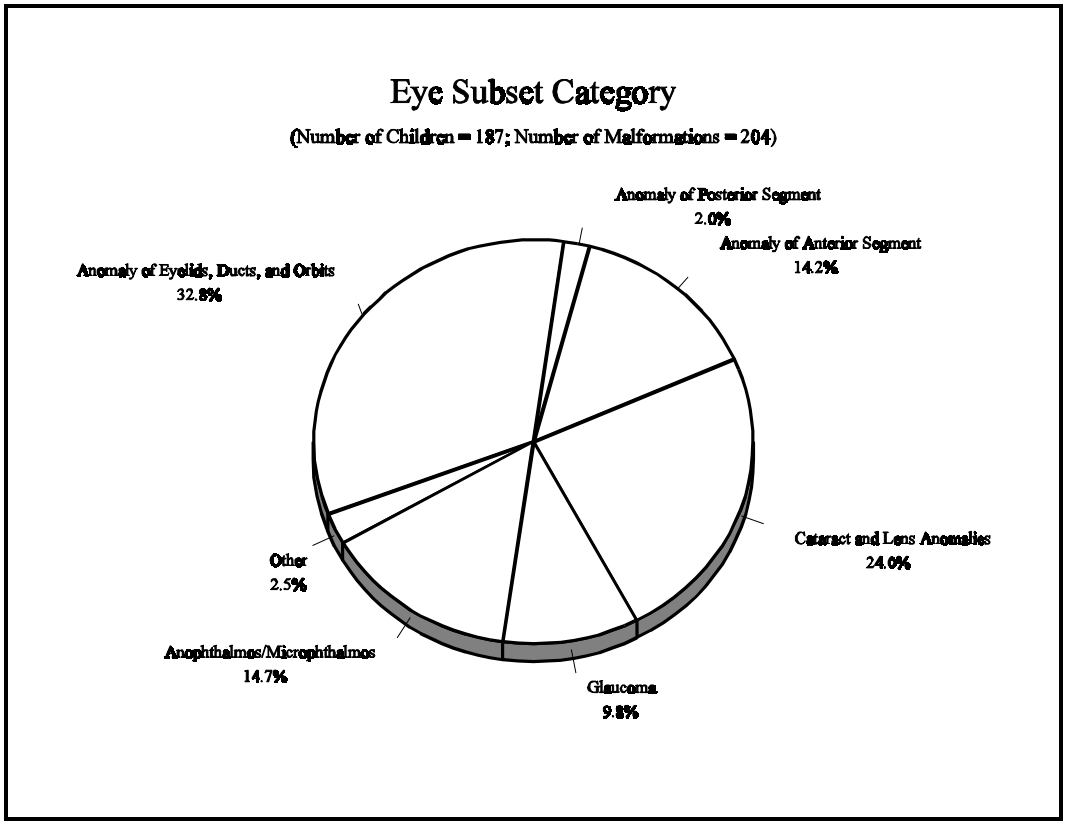
The organ system figures in this section present the distribution of 12 categories of major malformations, the relative contribution of each category to the overall incidence of major congenital malformations in New York State, and the contribution of type of malformation within each subset category.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

Major Malformations by Organ System
1994 Births - New York State Residents

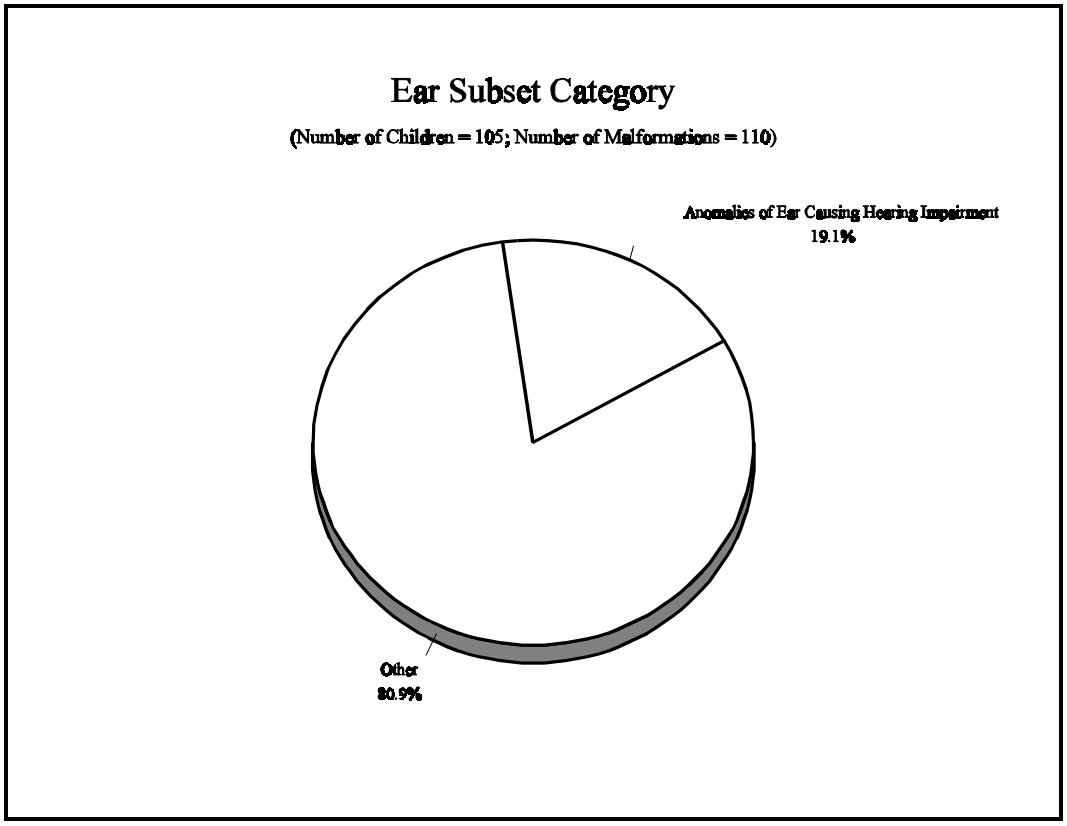


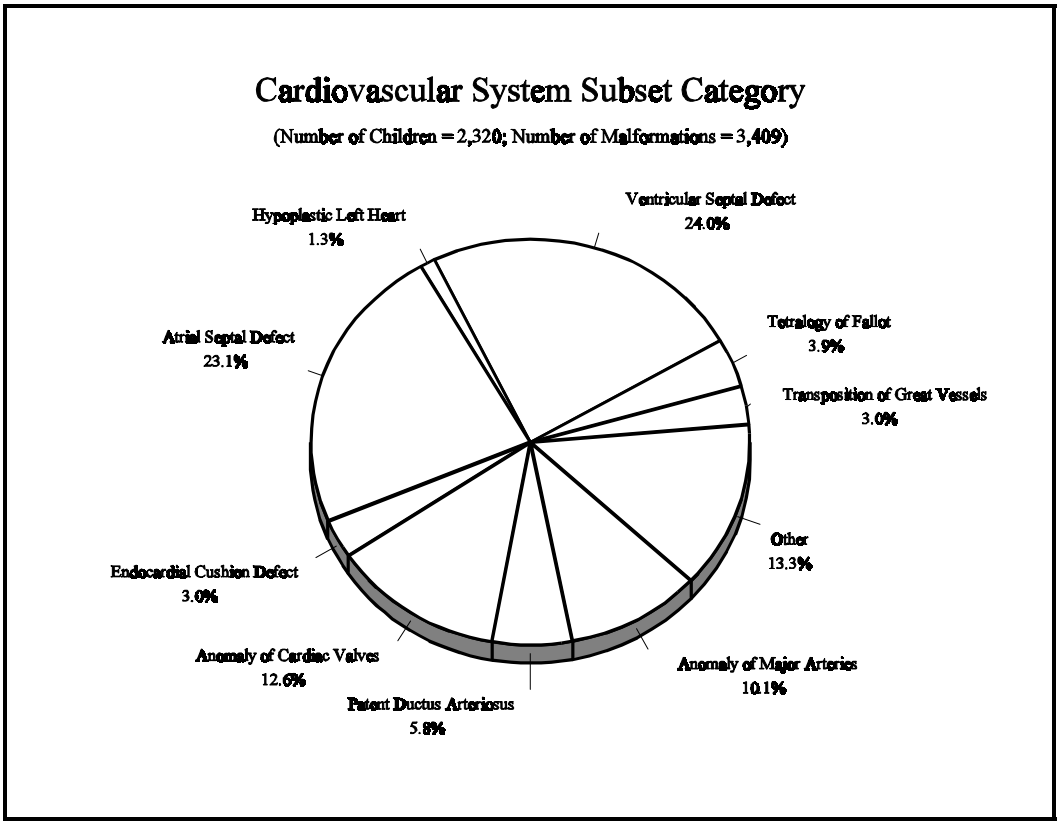
Major Malformations by Organ System
1994 Births - New York State Residents



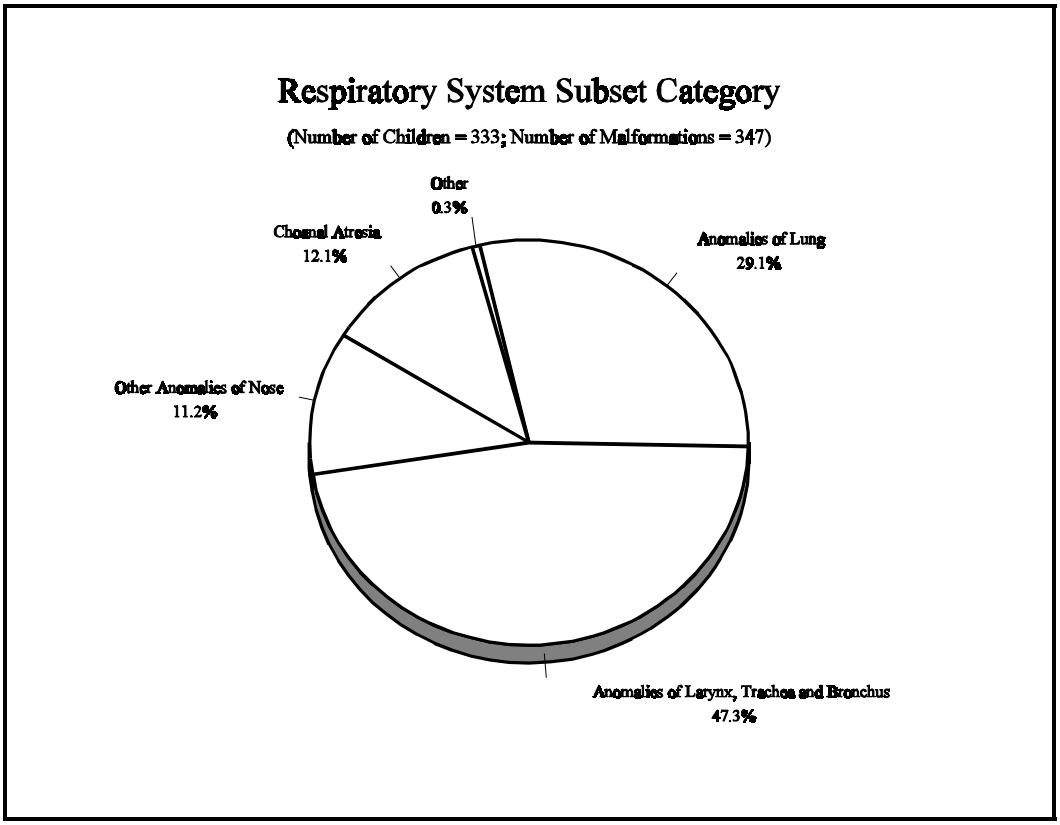


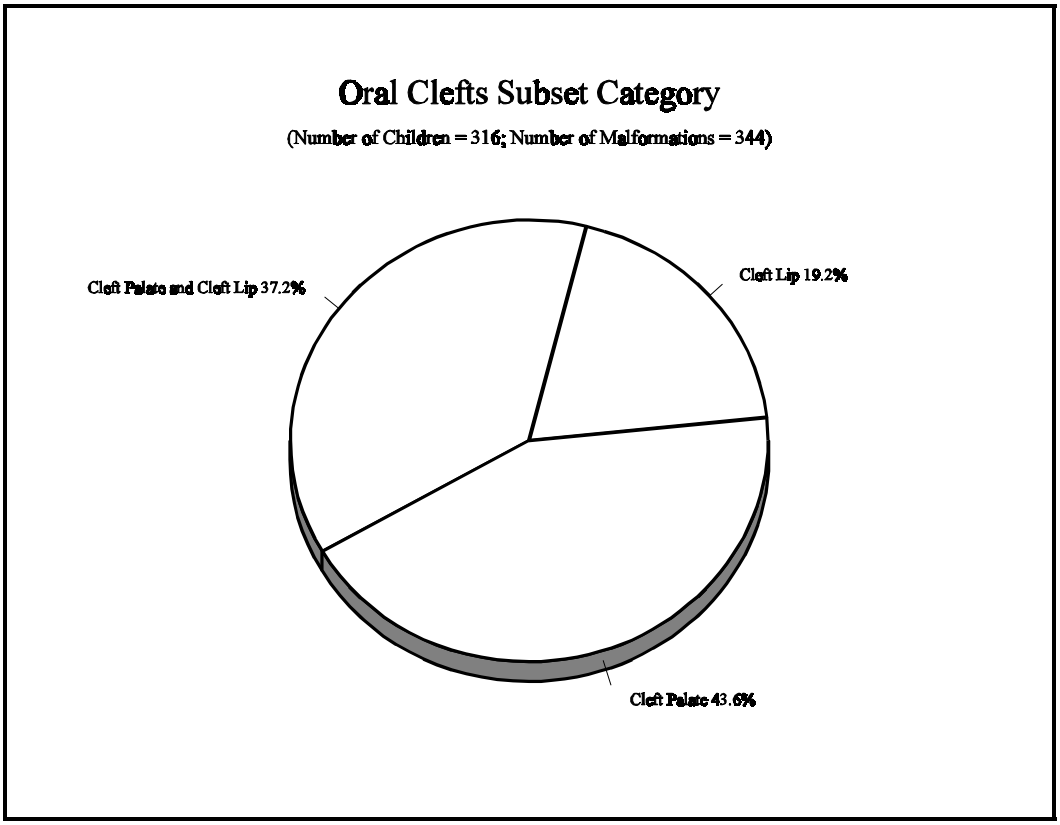
Major Malformations by Organ System
1994 Births - New York State Residents



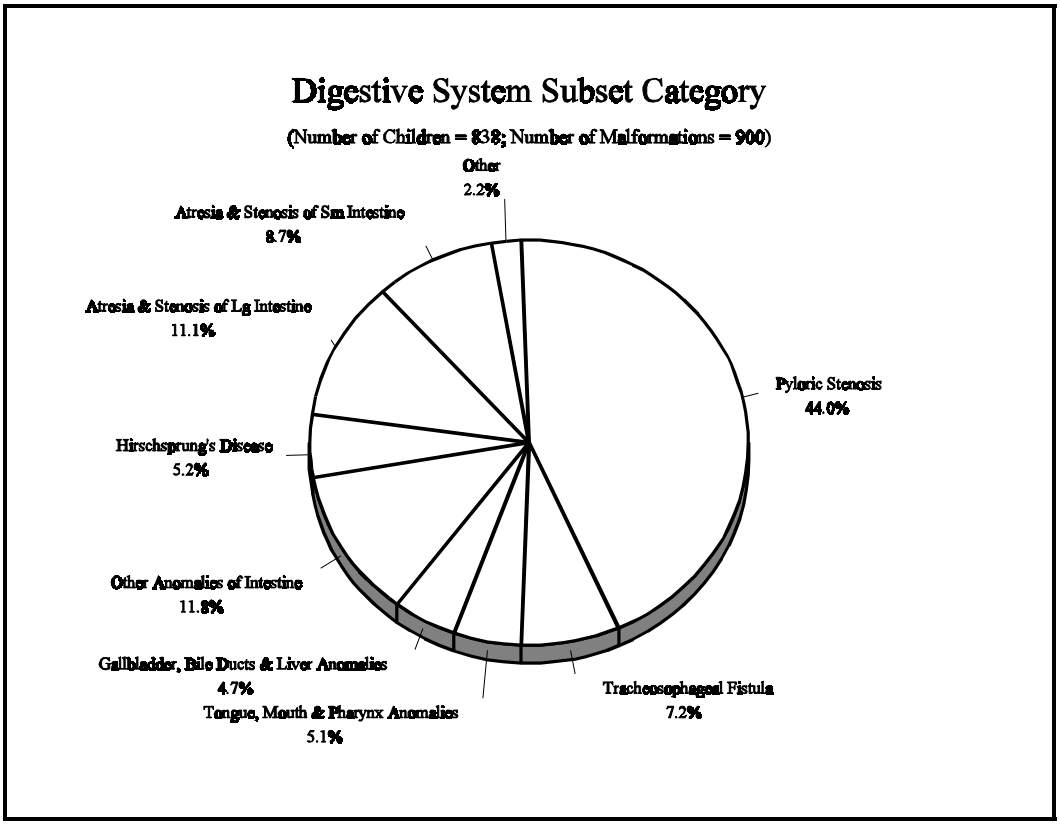


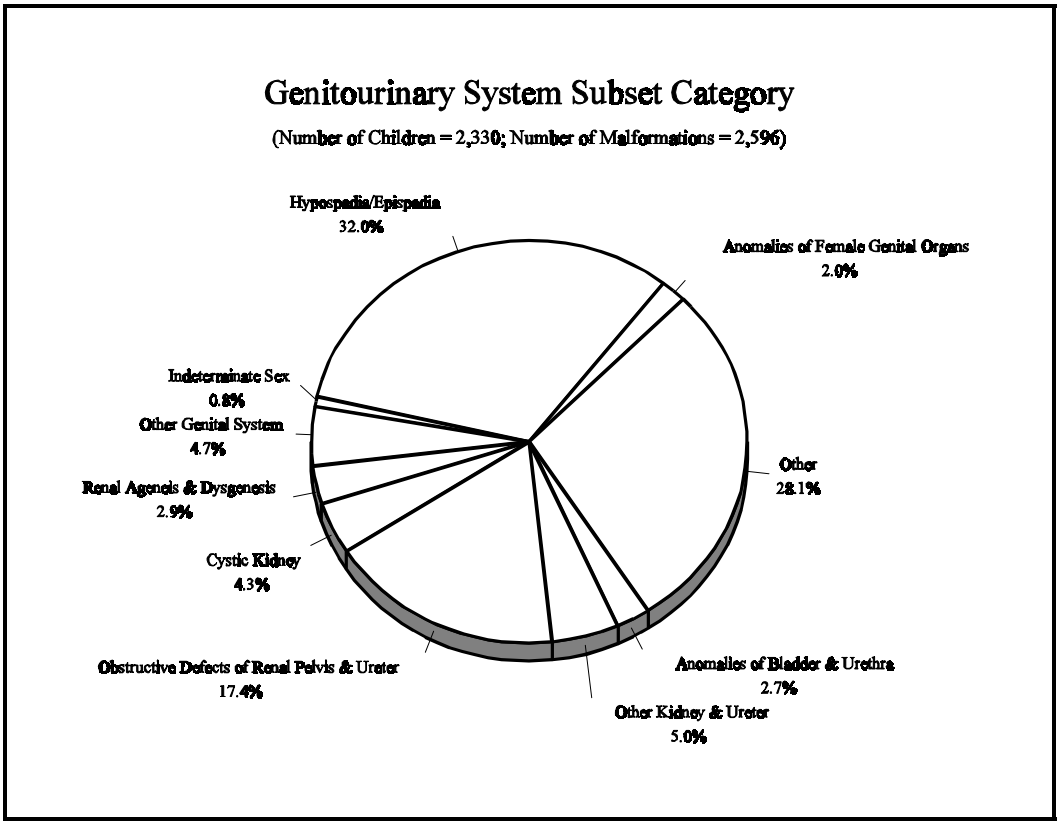
Major Malformations by Organ System
1994 Births - New York State Residents





Major Malformations by Organ System
1994 Births - New York State Residents

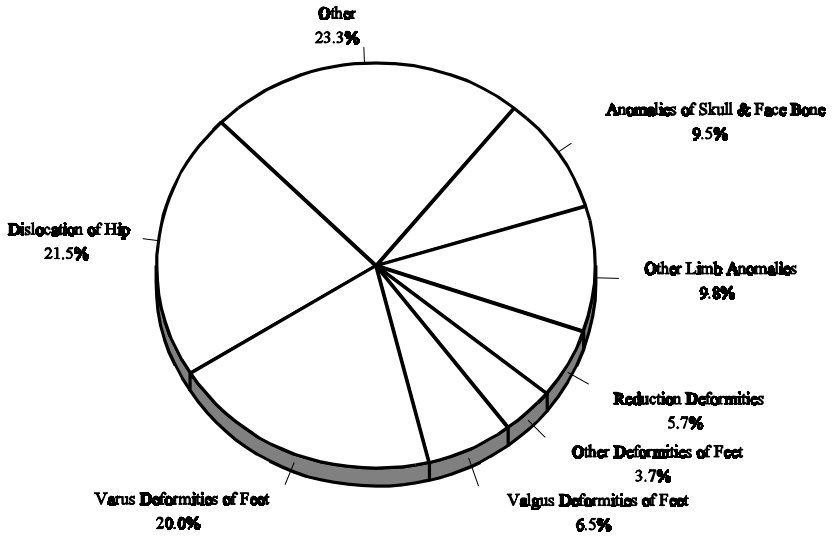




Major Malformations by Organ System
1994 Births - New York State Residents

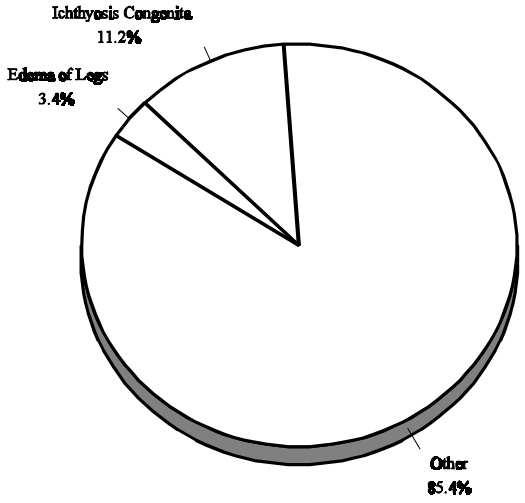
Musculoskeletal System Subset Category

(Number of Children = 2,045; Number of Malformations = 2,391)

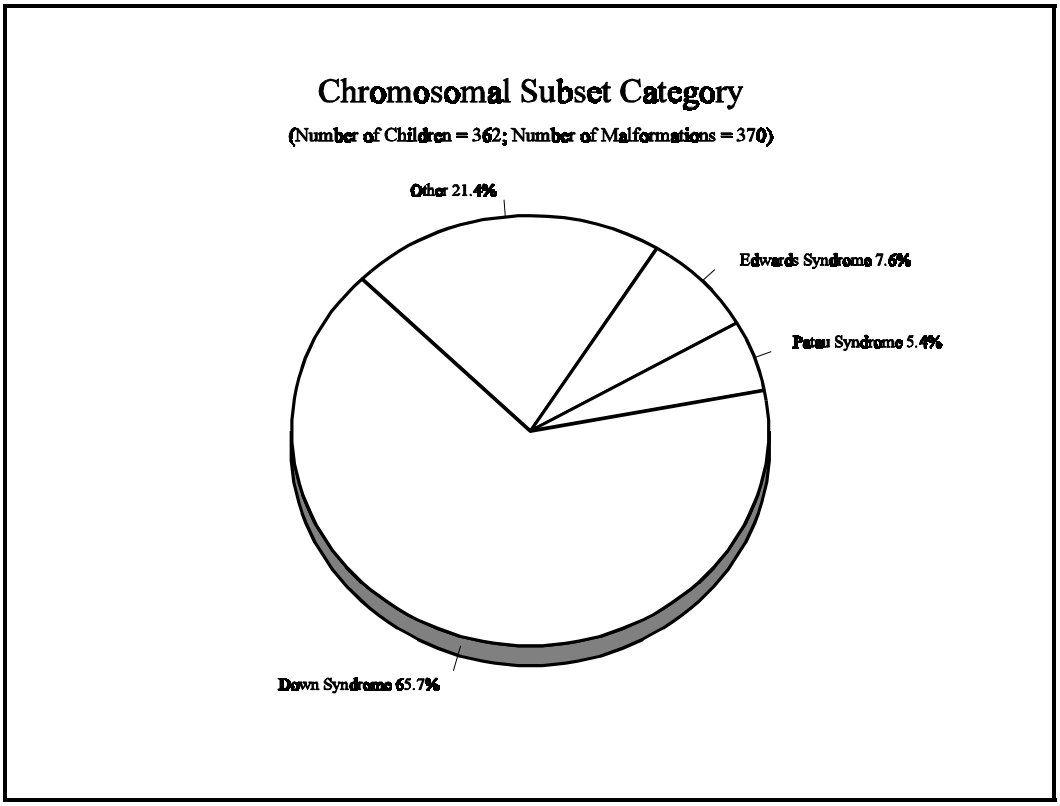


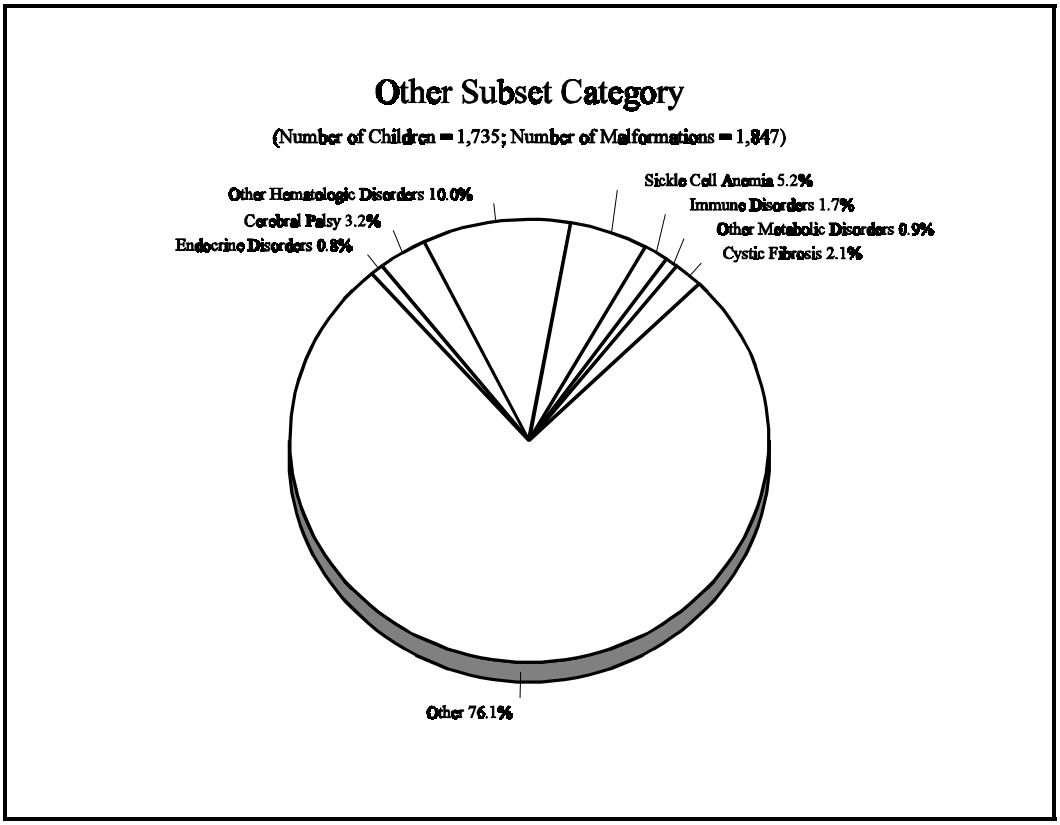
Integument Subset Category

(Number of Children = 89; Number of Malformations = 89)



Major Malformations by Organ System
1994 Births - New York State Residents





Section III
Prevalence of Selected Malformations
Rates 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
Prevalence per 10,000 Live Births by Sex & by Race

1994 Births - New York State Residents

ICD-9 Code	Malformation	Total Number	Total Prevalence	Male	Female	Ratio (M/F)	White	Black	Other
090	Congenital syphilis	584	21.0	21.2	20.8	1.0	9.9	64.3	4.5
243	Congenital hypothyroidism	48	1.7	2.0	1.4	1.5	1.4	2.8	2.6
270.1	Phenylketonuria	2	0.1	0.1	0.1	1.0	0.1	0.0	0.0
277.0	Cystic fibrosis	41	1.5	1.1	1.8	0.6	1.9	0.2	0.6
282.6	Sickle-cell anemia	108	3.9	4.3	3.5	1.2	0.6	16.2	0.6
658.8	Amniotic bands	9	0.3	0.3	0.4	0.8	0.2	0.7	0.0
740.0	Anencephalus	11	0.4	0.3	0.5	0.5	0.2	0.3	2.6
741.0	Spina bifida with hydrocephalus	64	2.3	2.3	2.3	1.0	2.7	1.6	0.0
741.9	Spina bifida without hydrocephalus	51	1.8	1.4	2.3	0.6	2.1	1.2	1.3
742.0	Encephalocele	20	0.7	0.6	0.9	0.6	0.7	1.0	0.0
742.1	Microcephalus	152	5.5	4.5	6.5	0.7	4.4	9.7	4.5
742.2	Agyria & lissencephaly	7	0.3	0.4	0.1	4.8	0.3	0.2	0.0
742.2	Anomalies of corpus callosum	27	1.0	1.0	0.9	1.1	0.9	1.0	0.6
742.2	Holoprosencephaly	11	0.4	0.4	0.4	1.2	0.3	0.7	0.0
742.3	Congenital hydrocephalus	218	7.8	8.8	6.8	1.3	7.1	10.5	7.1
742.4	Porencephaly	22	0.8	0.8	0.8	1.0	0.7	1.0	0.0
742.5	Congenital tethered cord	35	1.3	0.9	1.6	0.6	1.5	0.5	1.3
743.0	Anophthalmos	10	0.4	0.4	0.4	1.0	0.3	0.2	0.6
743.1	Microphthalmus	20	0.7	0.6	0.8	0.8	0.5	1.4	0.6
743.2	Glaucoma	21	0.8	0.8	0.7	1.3	0.7	1.0	0.0
743.3	Congenital cataract	49	1.8	1.6	2.0	0.8	1.9	1.6	0.6
743.45	Aniridia	5	0.2	0.1	0.3	0.2	0.1	0.5	0.0
743.46	Coloboma of iris	4	0.1	0.3	0.0	.	0.1	0.2	0.0
744.0	Ear anomalies with hearing impairment	22	0.8	0.8	0.8	1.0	0.9	0.3	1.3
745.0	Common truncus	21	0.8	0.8	0.7	1.1	0.6	1.0	1.3
745.1	Transposition of great vessels	115	4.1	4.7	3.5	1.3	4.1	4.0	5.2
745.2	Tetralogy of Fallot	132	4.7	4.7	4.9	1.0	4.6	4.5	7.7
745.3	Common ventricle	21	0.8	0.6	0.9	0.7	0.8	0.3	1.3
745.4	Ventricular septal defect	817	29.4	27.4	31.5	0.9	30.4	27.6	27.0
745.5	Ostium secundum type atrial septal def.	821	29.5	27.4	31.8	0.9	27.0	39.6	28.3
745.6	Endocardial cushion defects	121	4.4	3.9	4.9	0.8	4.4	4.5	3.9
746.0	Atresia/stenosis of pulmonary valve	232	8.3	7.8	8.9	0.9	7.1	13.3	6.4
746.1	Tricuspid atresia/stenosis/hypoplasia	29	1.0	1.1	1.0	1.0	1.0	0.9	1.3
746.2	Ebstein's anomaly	11	0.4	0.4	0.4	1.2	0.4	0.3	0.0
746.3	Congenital stenosis of aortic valve	51	1.8	1.8	1.9	0.9	2.0	1.6	0.6
746.7	Hypoplastic left heart syndrome	43	1.5	1.5	1.6	0.9	1.5	1.6	1.3
746.8	Anomalies of coronary artery	12	0.4	0.5	0.4	1.3	0.3	0.7	0.0
746.8	Dextrocardia without situs inversus	26	0.9	1.3	0.6	2.2	0.7	2.1	0.0
747.0	Patent ductus arteriosis	201	7.2	7.1	7.4	1.0	6.8	8.5	7.7
747.1	Coarctation of aorta	73	2.6	2.8	2.4	1.2	3.0	1.2	3.9
747.3	Anomalies of pulmonary artery	229	8.2	7.3	9.3	0.8	7.0	13.3	7.1
747.41	Total anomalous pulmonary venus connect.	16	0.6	0.8	0.4	2.1	0.4	1.2	0.0

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

1994 Births - New York State Residents

ICD-9 Code	Malformation	Total Number	Total Prevalence	Male	Female	Ratio (M/F)	White	Black	Other
748.0	Choanal atresia	42	1.5	1.3	1.8	0.7	1.7	0.9	0.6
749.0	Cleft palate	176	6.3	5.1	7.6	0.7	7.2	3.8	4.5
749.1	Cleft lip	77	2.8	3.5	2.1	1.7	2.6	1.6	9.0
749.2	Cleft palate & lip	172	6.2	7.2	5.1	1.4	7.2	2.4	8.4
750.3	Tracheoesophageal fistula, etc.	71	2.6	2.9	2.2	1.3	2.8	1.7	3.2
750.5	Congenital hypertrophic pyloric stenosis	396	14.2	22.5	5.6	4.0	16.4	8.6	7.7
751.1	Atresia and stenosis of duodenum	29	1.0	1.1	1.0	1.0	0.9	1.9	0.0
751.1	Atresia and stenosis of small intestine	60	2.2	1.8	2.5	0.7	1.6	4.0	2.6
751.2	Atresia and stenosis of rectum or anus	79	2.8	3.2	2.5	1.3	2.9	2.4	3.9
751.3	Hirschsprung's disease	44	1.6	2.0	1.1	1.9	1.6	1.4	1.9
751.4	Anomalies of intestinal fixation	47	1.7	1.3	2.1	0.6	1.5	2.1	2.6
751.61	Biliary atresia	12	0.4	0.3	0.6	0.5	0.4	0.3	0.6
752.6	Epispadias	16	0.6	1.0	0.1	6.7	0.5	0.9	0.0
752.6	Hypospadias	857	30.8	59.7	0.6	101.5	34.2	21.6	25.1
753.0	Renal agenesis and dysgenesis	79	2.8	3.5	2.1	1.7	2.6	2.9	6.4
753.1	Cystic kidney disease	81	2.9	3.3	2.5	1.3	2.7	3.8	2.6
753.2	Obstructive defect renal pelvis & ureter	509	18.3	25.2	11.1	2.3	19.7	13.6	19.3
753.5	Extrophy of urinary bladder	8	0.3	0.3	0.3	1.0	0.2	0.3	0.6
753.6	Atresia & stenosis of urethra & bladder	38	1.4	2.5	0.2	11.2	1.5	1.0	0.6
754.2	Musculoskeletal deformities of spine	17	0.6	0.7	0.5	1.4	0.5	1.0	0.6
754.3	Congenital dislocation of hip	504	18.1	9.4	27.2	0.3	20.9	7.9	22.5
754.5	Talipes equinovarus	282	10.1	12.0	8.2	1.5	10.4	9.8	9.0
755.2	Reduction deformities of upper limb	92	3.3	2.6	4.0	0.6	3.5	3.3	1.3
755.3	Reduction deformities of lower limb	79	2.8	3.4	2.1	1.6	2.7	3.5	1.9
755.8	Arthrogryposis multiplex congenita	43	1.5	1.6	1.5	1.1	1.5	1.9	1.3
756.0	Craniosynostosis	118	4.2	5.4	3.1	1.7	5.1	1.0	5.8
756.0	Goldenhar syndrome	12	0.4	0.7	0.1	4.8	0.4	0.5	0.6
756.4	Chondrodystrophy	22	0.8	0.7	0.9	0.8	0.8	1.0	0.0
756.5	Osteogenesis imperfecta	17	0.6	0.8	0.4	1.8	0.4	1.2	0.6
756.6	Diaphragmatic hernia	58	2.1	2.5	1.6	1.6	2.1	1.4	4.5
756.7	Gastroschisis	37	1.3	1.6	1.1	1.4	1.8	0.2	0.0
756.7	Omphalocele	36	1.3	1.4	1.2	1.2	1.2	2.1	0.0
756.7	Prune belly	9	0.3	0.5	0.1	3.4	0.3	0.3	0.0
758.0	Down syndrome	243	8.7	9.0	8.5	1.1	9.4	7.8	4.5
758.1	Patau syndrome	21	0.8	0.8	0.7	1.3	0.7	1.0	0.6
758.2	Edwards syndrome	28	1.0	0.5	1.5	0.3	1.0	1.2	0.0
758.6	Gonadal dysgenesis	12	0.4	0.0	0.9	0.0	0.5	0.3	0.0
758.7	Klinefelter syndrome	10	0.4	0.7	0.0	.	0.3	0.3	1.3
759.3	Situs inversus	23	0.8	0.8	0.8	1.0	0.7	1.2	0.6
760.71	Fetal alcohol syndrome	56	2.0	2.3	1.6	1.4	0.9	6.0	1.3
771.0	Congenital rubella	2	0.1	0.1	0.1	1.0	0.0	0.2	0.0
771.1	Congenital cytomegalovirus infection	27	1.0	0.8	1.1	0.8	0.4	2.6	1.3
771.2	Other congenital infections	26	0.9	0.9	1.0	1.0	0.6	2.2	0.0

Section IV
Most Frequently Reported Selected Major Malformations by County

Introduction to Tables

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

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.

Since many factors affect reporting, these county listings should not be used for comparisons or analytic studies. They may be most useful to assist in county planning, education, counseling and other health care service programs.

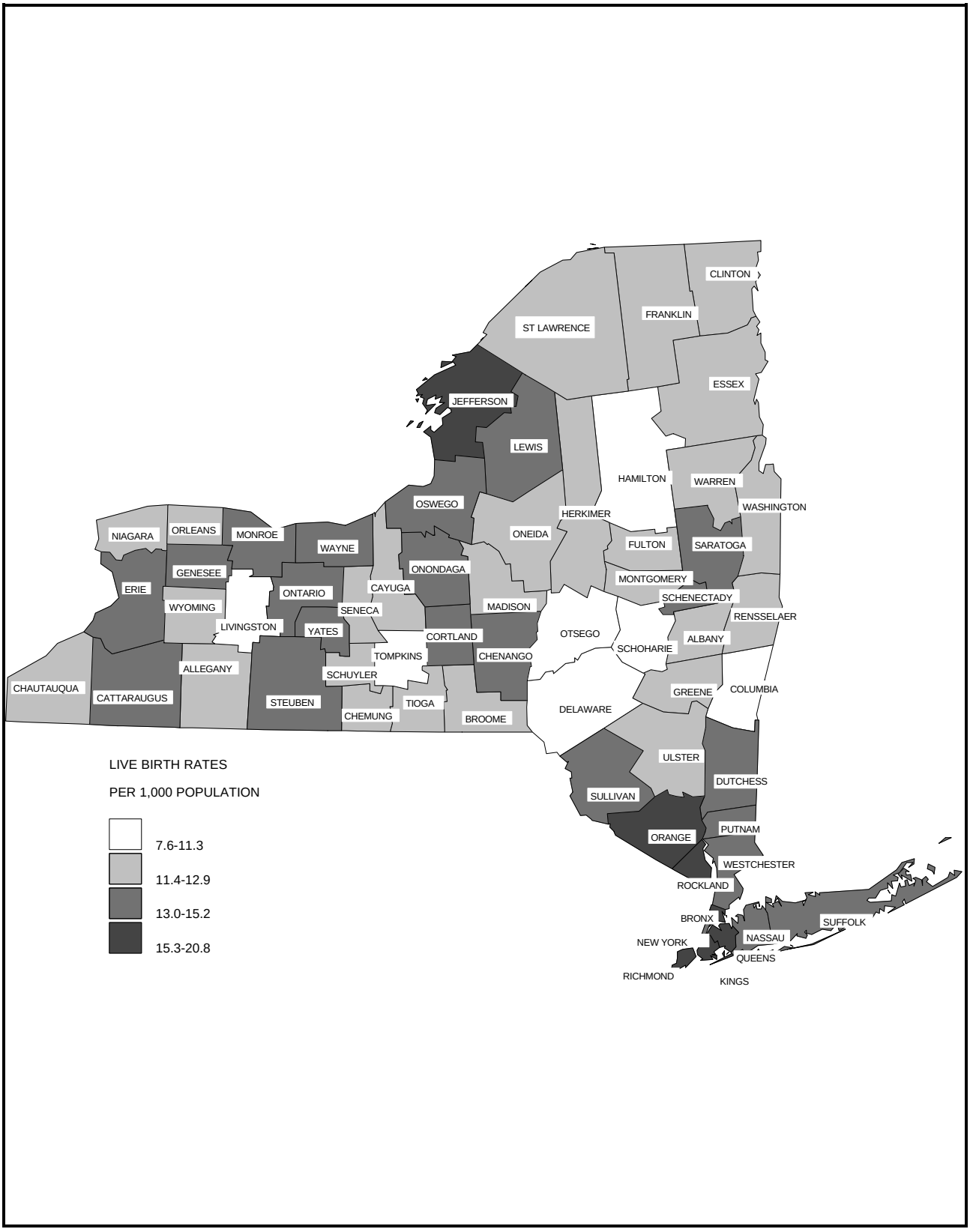
The map of New York State showing county live birth rates is part of the Bureau of Biometrics annual report, "Vital Statistics of New York State 1994." The Bureau of Biometrics supplies the CMR with the number of live births in a specific year, which is used as a denominator in the calculation of prevalences of malformations. For 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, 1994

County	Number of Children	Number of Live Births	Percent of Live Births
Albany	118	3,539	3.3
Allegany	11	608	1.8
Bronx	843	25,453	3.3
Broome	71	2,474	2.9
Cattaraugus	36	1,100	3.3
Cayuga	44	1,016	4.3
Chautauqua	53	1,756	3.0
Chemung	46	1,156	4.0
Chenango	33	687	4.8
Clinton	21	1,054	2.0
Columbia	17	726	2.3
Cortland	21	655	3.2
Delaware	10	536	1.9
Dutchess	80	3,452	2.3
Erie	567	12,850	4.4
Essex	7	459	1.5
Franklin	7	615	1.1
Fulton	45	701	6.4
Genesee	39	859	4.5
Greene	8	542	1.5
Hamilton	1	41	2.4
Herkimer	29	820	3.5
Jefferson	48	1,910	2.5
Kings	1,546	43,413	3.6
Lewis	10	382	2.6
Livingston	28	734	3.8
Madison	36	897	4.0
Monroe	363	10,500	3.5
Montgomery	23	672	3.4
Nassau	720	17,903	4.0
New York	759	21,331	3.6
Niagara	97	2,909	3.3
Oneida	101	3,134	3.2
Onondaga	283	6,752	4.2
Ontario	63	1,275	4.9
Orange	150	5,030	3.0
Orleans	31	578	5.4
Oswego	93	1,721	5.4
Otsego	20	662	3.0
Putnam	25	1,275	2.0
Queens	1,145	33,025	3.5
Rensselaer	78	2,018	3.9
Richmond	195	6,105	3.2
Rockland	98	4,279	2.3
St. Lawrence	18	1,335	1.3
Saratoga	102	2,619	3.9
Schenectady	59	2,035	2.9
Schoharie	10	379	2.6
Schuyler	10	248	4.0
Seneca	13	397	3.3
Steuben	45	1,346	3.3
Suffolk	550	20,502	2.7
Sullivan	22	972	2.3
Tioga	17	667	2.5
Tompkins	28	1,035	2.7
Ulster	68	2,143	3.2
Warren	31	788	3.9
Washington	14	719	1.9
Wayne	42	1,329	3.2
Westchester	377	13,002	2.9
Wyoming	25	509	4.9
Yates	5	316	1.6

Live Birth Rates, New York State 1994



Section IV
Ten Most Frequently Reported Major Malformations
By County, 1994
Congenital Malformations Registry

County	ICD - 9 Code	Description	Number Reported
Albany			
	745.4	Ventricular septal defect	13
	745.5	Ostium secundum atrial septal defect	13
	752.6	Hypospadias & epispadias	11
	746.8	Other specified anomalies of heart	7
	753.2	Obstructive defects of renal pelvis & ureter	6
	754.3	Congenital dislocation of hip	6
	750.5	Congenital hypertrophic pyloric stenosis	5
	753.4	Other specified anomalies of ureter	5
	752.5	Undescended testicle	4
	758.0	Down syndrome	4
Allegany			
	752.5	Undescended testicle	2
	741.9	Spina bifida w/o hydrocephalus	1
	745.4	Ventricular septal defect	1
	747.3	Anomalies of pulmonary artery	1
	749.2	Cleft palate with cleft lip	1
	753.2	Obstructive defects of renal pelvis & ureter	1
	754.5	Varus deformities of feet	1
	758.0	Down syndrome	1
Bronx			
	752.6	Hypospadias & epispadias	67
	752.5	Undescended testicle	66
	745.5	Ostium secundum atrial septal defect	53
	745.4	Ventricular septal defect	51
	754.5	Varus deformities of feet	46
	754.3	Congenital dislocation of hip	26
	750.5	Congenital hypertrophic pyloric stenosis	23
	758.0	Down syndrome	22
	747.3	Anomalies of pulmonary artery	19
	748.3	Other anomalies of larynx, trachea, & bronchus	19
	753.2	Obstructive defects of renal pelvis & ureter	19
	756.0	Anomalies of skull and face bones	19
Broome			
	752.5	Undescended testicle	10
	745.5	Ostium secundum atrial septal defect	9
	754.3	Congenital dislocation of hip	9
	752.6	Hypospadias & epispadias	6
	754.5	Varus deformities of feet	5
	745.4	Ventricular septal defect	4
	746.0	Anomalies of pulmonary valve	3
	750.5	Congenital hypertrophic pyloric stenosis	3
	742.3	Congenital hydrocephalus	2
	745.2	Tetralogy of Fallot	2
	746.8	Other specified anomalies of heart	2
	747.3	Anomalies of pulmonary artery	2
	748.5	Agenesis, hypoplasia & dysplasia, lung	2
	749.1	Cleft lip	2
	749.2	Cleft palate with cleft lip	2

Section IV
Ten Most Frequently Reported Major Malformations
By County, 1994
Congenital Malformations Registry

County	ICD - 9 Code	Description	Number Reported
Broome, cont'd.			
	751.2	Atresia and stenosis of large intestine, rectum, & anal canal	2
	753.1	Cystic kidney disease	2
	755.6	Other anomalies of lower limb including pelvic girdle	2
	756.0	Anomalies of skull and face bones	2
Cattaraugus			
	752.6	Hypospadias & epispadias	6
	745.4	Ventricular septal defect	4
	745.5	Ostium secundum atrial septal defect	4
	747.3	Anomalies of pulmonary artery	3
	743.3	Congenital cataract & lens anomalies	2
	746.0	Anomalies of pulmonary valve	2
	747.2	Other anomalies of aorta	2
	748.3	Other anomalies of larynx, trachea, & bronchus	2
	751.2	Atresia and stenosis of large intestine, rectum, & anal canal	2
	751.3	Hirschprung's disease & other functional disorders of colon	2
	756.0	Anomalies of skull and face bones	2
	758.0	Down syndrome	2
Cayuga			
	752.5	Undescended testicle	9
	752.6	Hypospadias & epispadias	9
	745.5	Ostium secundum atrial septal defect	5
	745.4	Ventricular septal defect	3
	750.5	Congenital hypertrophic pyloric stenosis	3
	753.2	Obstructive defects of renal pelvis & ureter	3
	755.6	Other anomalies of lower limb including pelvic girdle	3
	746.0	Anomalies of pulmonary valve	2
	746.1	Tricuspid atresia & stenosis	2
	746.8	Other specified anomalies of heart	2
	747.3	Anomalies of pulmonary artery	2
	752.8	Other specified anomalies of genital organs	2
Chautauqua			
	745.4	Ventricular septal defect	10
	745.5	Ostium secundum atrial septal defect	8
	752.5	Undescended testicle	8
	748.3	Other anomalies of larynx, trachea, & bronchus	4
	746.0	Anomalies of pulmonary valve	3
	746.8	Other specified anomalies of heart	3
	754.5	Varus deformities of feet	3
	758.0	Down syndrome	3
	745.1	Transposition of great vessels	2
	746.6	Congenital mitral insufficiency	2
	748.5	Agenesis, hypoplasia & dysplasia, lung	2
	750.5	Congenital hypertrophic pyloric stenosis	2
	752.6	Hypospadias & epispadias	2
	753.2	Obstructive defects of renal pelvis & ureter	2
	756.5	Osteodystrophies	2
Chemung			
	750.5	Congenital hypertrophic pyloric stenosis	5

Section IV
Ten Most Frequently Reported Major Malformations
By County, 1994
Congenital Malformations Registry

County	ICD - 9 Code	Description	Number Reported
Chemung cont'd			
	745.4	Ventricular septal defect	4
	752.5	Undescended testicle	4
	745.5	Ostium secundum atrial septal defect	3
	752.6	Hypospadias & epispadias	3
	746.0	Anomalies of pulmonary valve	2
	746.8	Other specified anomalies of heart	2
	749.2	Cleft palate with cleft lip	2
	756.0	Anomalies of skull and face bones	2
	352.6	Multiple cranial nerve palsies	1
	740.0	Anencephalus	1
	742.5	Other specified anomalies of spinal cord	1
	745.6	Endocardial cushion defects	1
	746.4	Congenital insufficiency of aortic arch	1
	746.6	Congenital mitral insufficiency	1
	747.1	Coarctation of aorta	1
	747.2	Other anomalies of aorta	1
	749.1	Cleft lip	1
	750.3	Tracheoesophageal fistula, esophageal atresia & stenosis	1
	751.4	Anomalies of intestinal fixation	1
	753.2	Obstructive defects of renal pelvis & ureter	1
	754.3	Congenital dislocation of hip	1
	754.5	Varus deformities of feet	1
	756.6	Anomalies of diaphragm	1
	756.7	Anomalies of abdominal wall	1
	757.3	Other specified anomalies of skin	1
	758.0	Down syndrome	1
	758.6	Gonadal dysgenesis	1
	759.3	Situs inversus	1
Chenango			
	754.3	Congenital dislocation of hip	7
	745.5	Ostium secundum atrial septal defect	6
	752.5	Undescended testicle	3
	754.5	Varus deformities of feet	3
	749.2	Cleft palate with cleft lip	2
	755.6	Other anomalies of lower limb including pelvic girdle	2
	742.1	Microcephalus	1
	742.2	Reduction deformities of brain	1
	742.3	Congenital hydrocephalus	1
	745.0	Common truncus	1
	745.4	Ventricular septal defect	1
	746.6	Congenital mitral insufficiency	1
	746.7	Hypoplastic left heart syndrome	1
	746.8	Other specified anomalies of heart	1
	747.0	Patent ductus arteriosus	1
	747.1	Coarctation of aorta	1
	748.3	Other anomalies of larynx, trachea, & bronchus	1
	749.0	Cleft palate	1
	750.5	Congenital hypertrophic pyloric stenosis	1
	751.2	Atresia and stenosis of large intestine, rectum, & anal canal	1
	752.6	Hypospadias & epispadias	1

Section IV
Ten Most Frequently Reported Major Malformations
By County, 1994
Congenital Malformations Registry

County	ICD - 9 Code	Description	Number Reported
Chenango, cont'd.			
	753.2	Obstructive defects of renal pelvis & ureter	1
	753.8	Other specified anomalies of bladder & urethra	1
	756.6	Anomalies of diaphragm	1
Clinton			
	752.5	Undescended testicle	4
	745.4	Ventricular septal defect	2
	749.0	Cleft palate	2
	752.6	Hypospadias & epispadias	2
	753.2	Obstructive defects of renal pelvis & ureter	2
	277.0	Cystic fibrosis	1
	740.0	Anencephalus	1
	743.3	Congenital cataract & lens anomalies	1
	749.2	Cleft palate with cleft lip	1
	750.5	Congenital hypertrophic pyloric stenosis	1
	751.2	Atresia and stenosis of large intestine, rectum, & anal canal	1
	752.0	Anomalies of ovaries	1
	752.8	Other specified anomalies of genital organs	1
	753.1	Cystic kidney disease	1
	754.5	Varus deformities of feet	1
	755.6	Other anomalies of lower limb including pelvic girdle	1
	756.7	Anomalies of abdominal wall	1
Columbia			
	277.0	Cystic fibrosis	2
	742.3	Congenital hydrocephalus	2
	745.4	Ventricular septal defect	2
	745.5	Ostium secundum atrial septal defect	2
	754.5	Varus deformities of feet	2
	282.0	Hereditary spherocytosis	1
	742.2	Reduction deformities of brain	1
	742.4	Other specified anomalies of brain	1
	744.0	Anomalies of ear causing impairment of hearing	1
	746.0	Anomalies of pulmonary valve	1
	746.8	Other specified anomalies of heart	1
	750.5	Congenital hypertrophic pyloric stenosis	1
	751.2	Atresia and stenosis of large intestine, rectum, & anal canal	1
	752.6	Hypospadias & epispadias	1
	753.0	Renal agenesis & dysgenesis	1
	753.2	Obstructive defects of renal pelvis & ureter	1
	753.4	Other specified anomalies of ureter	1
	754.3	Congenital dislocation of hip	1
	756.5	Osteodystrophies	1
	758.0	Down syndrome	1
	758.2	Edwards syndrome	1
Cortland			
	745.5	Ostium secundum atrial septal defect	3
	748.3	Other anomalies of larynx, trachea, & bronchus	2
	749.0	Cleft palate	2

Section IV
Ten Most Frequently Reported Major Malformations
By County, 1994
Congenital Malformations Registry

County	ICD - 9 Code	Description	Number Reported
Cortland cont'd			
	752.6	Hypospadias & epispadias	2
	756.7	Anomalies of abdominal wall	2
	742.1	Microcephalus	1
	742.2	Reduction deformities of brain	1
	742.3	Congenital hydrocephalus	1
	742.4	Other specified anomalies of brain	1
	747.0	Patent ductus arteriosus	1
	747.3	Anomalies of pulmonary artery	1
	748.0	Choanal atresia	1
	750.5	Congenital hypertrophic pyloric stenosis	1
	751.2	Atresia and stenosis of large intestine, rectum, & anal canal	1
	754.5	Varus deformities of feet	1
	756.0	Anomalies of skull and face bones	1
	758.0	Down syndrome	1
	758.1	Patau syndrome	1
Delaware			
	745.5	Ostium secundum atrial septal defect	3
	742.0	Encephalocele	1
	742.1	Microcephalus	1
	742.3	Congenital hydrocephalus	1
	747.0	Patent ductus arteriosus	1
	749.0	Cleft palate	1
	750.5	Congenital hypertrophic pyloric stenosis	1
	751.1	Atresia and stenosis of small intestine	1
	753.1	Cystic kidney disease	1
	753.6	Atresia and stenosis of urethra & bladder neck	1
	755.2	Reduction deformities of upper limb	1
	755.6	Other anomalies of lower limb including pelvic girdle	1
	756.7	Anomalies of abdominal wall	1
	758.0	Down syndrome	1
	758.2	Edwards syndrome	1
	758.6	Gonadal dysgenesis	1
Dutchess			
	752.6	Hypospadias & epispadias	14
	745.4	Ventricular septal defect	9
	745.5	Ostium secundum atrial septal defect	8
	752.5	Undescended testicle	7
	746.8	Other specified anomalies of heart	5
	754.5	Varus deformities of feet	5
	758.0	Down syndrome	5
	742.1	Microcephalus	4
	753.2	Obstructive defects of renal pelvis & ureter	4
	754.3	Congenital dislocation of hip	4
	756.0	Anomalies of skull and face bones	4
Erie			
	752.6	Hypospadias & epispadias	70
	745.4	Ventricular septal defect	57
	752.5	Undescended testicle	51

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County	ICD - 9 Code	Description	Number Reported
Erie cont'd			
	754.5	Varus deformities of feet	39
	750.5	Congenital hypertrophic pyloric stenosis	28
	747.3	Anomalies of pulmonary artery	27
	745.5	Ostium secundum atrial septal defect	26
	754.3	Congenital dislocation of hip	26
	746.8	Other specified anomalies of heart	19
	747.0	Patent ductus arteriosus	18
	752.8	Other specified anomalies of genital organs	18
Essex			
	745.5	Ostium secundum atrial septal defect	2
	746.8	Other specified anomalies of heart	2
	277.0	Cystic fibrosis	1
	740.0	Anencephalus	1
	742.1	Microcephalus	1
	745.4	Ventricular septal defect	1
	745.6	Endocardial cushion defects	1
	746.5	Congenital mitral stenosis	1
	746.6	Congenital mitral insufficiency	1
	747.1	Coarctation of aorta	1
	752.5	Undescended testicle	1
	753.2	Obstructive defects of renal pelvis & ureter	1
	753.4	Other specified anomalies of ureter	1
	758.0	Down syndrome	1
Franklin			
	754.3	Congenital dislocation of hip	2
	758.0	Down syndrome	2
	745.5	Ostium secundum atrial septal defect	1
	745.6	Endocardial cushion defects	1
	746.0	Anomalies of pulmonary valve	1
	752.7	Indeterminate sex & pseudo-hermaphroditism	1
	753.7	Anomalies of urachus	1
	754.5	Varus deformities of feet	1
	755.1	Syndactyly	1
	755.2	Reduction deformities of upper limb	1
	756.7	Anomalies of abdominal wall	1
Fulton			
	752.5	Undescended testicle	7
	746.8	Other specified anomalies of heart	6
	754.5	Varus deformities of feet	6
	745.4	Ventricular septal defect	4
	749.0	Cleft palate	3
	752.6	Hypospadias & epispadias	3
	745.5	Ostium secundum atrial septal defect	2
	756.0	Anomalies of skull and face bones	2
	742.1	Microcephalus	1
	743.2	Buphthalmos	1
	743.4	Coloboma & other anomalies of anterior segment	1
	744.0	Anomalies of ear causing impairment of hearing	1

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Fulton, cont'd.			
	745.2	Tetralogy of Fallot	1
	746.0	Anomalies of pulmonary valve	1
	746.6	Congenital mitral insufficiency	1
	747.0	Patent ductus arteriosus	1
	747.1	Coarctation of aorta	1
	747.2	Other anomalies of aorta	1
	747.3	Anomalies of pulmonary artery	1
	748.5	Agenesis, hypoplasia & dysplasia, lung	1
	749.2	Cleft palate with cleft lip	1
	750.3	Tracheoesophageal fistula, esophageal atresia & stenosis	1
	750.5	Congenital hypertrophic pyloric stenosis	1
	751.2	Atresia and stenosis of large intestine, rectum, & anal canal	1
	751.4	Anomalies of intestinal fixation	1
	752.8	Other specified anomalies of genital organs	1
	753.0	Renal agenesis & dysgenesis	1
	753.2	Obstructive defects of renal pelvis & ureter	1
	753.3	Other specified anomalies of kidney	1
	754.3	Congenital dislocation of hip	1
	755.1	Syndactyly	1
	755.5	Other anomalies of upper limb including shoulder girdle	1
	756.1	Anomalies of spine	1
	758.0	Down syndrome	1
Genesee			
	752.6	Hypospadias & epispadias	7
	750.5	Congenital hypertrophic pyloric stenosis	6
	752.5	Undescended testicle	5
	745.5	Ostium secundum atrial septal defect	3
	747.3	Anomalies of pulmonary artery	3
	745.4	Ventricular septal defect	2
	754.3	Congenital dislocation of hip	2
	755.2	Reduction deformities of upper limb	2
	755.6	Other anomalies of lower limb including pelvic girdle	2
	228.1	Lymphangioma, any site	1
	742.3	Congenital hydrocephalus	1
	745.1	Transposition of great vessels	1
	745.6	Endocardial cushion defects	1
	746.6	Congenital mitral insufficiency	1
	746.7	Hypoplastic left heart syndrome	1
	746.8	Other specified anomalies of heart	1
	748.3	Other anomalies of larynx, trachea, & bronchus	1
	749.1	Cleft lip	1
	749.2	Cleft palate with cleft lip	1
	750.3	Tracheoesophageal fistula, esophageal atresia & stenosis	1
	751.2	Atresia and stenosis of large intestine, rectum, & anal canal	1
	753.0	Renal agenesis & dysgenesis	1
	753.1	Cystic kidney disease	1
	753.2	Obstructive defects of renal pelvis & ureter	1
	754.5	Varus deformities of feet	1
	755.3	Reduction deformities of lower limb	1
	756.0	Anomalies of skull and face bones	1

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Genessee, cont'd.			
	756.6	Anomalies of diaphragm	1
	756.8	Other specified anomalies of muscle, tendon, fascia, etc.	1
	757.1	Ichthyosis congenita	1
	757.3	Other specified anomalies of skin	1
	758.2	Edwards syndrome	1
	759.8	Other specified anomalies	1
Greene			
	743.3	Congenital cataract & lens anomalies	2
	745.5	Ostium secundum atrial septal defect	2
	275.4	Disorders of calcium metabolism	1
	745.1	Transposition of great vessels	1
	746.0	Anomalies of pulmonary valve	1
	746.1	Tricuspid atresia & stenosis	1
	747.3	Anomalies of pulmonary artery	1
	748.3	Other anomalies of larynx, trachea, & bronchus	1
	753.2	Obstructive defects of renal pelvis & ureter	1
	753.4	Other specified anomalies of ureter	1
	756.0	Anomalies of skull and face bones	1
	758.0	Down syndrome	1
Hamilton			
	749.0	Cleft palate	1
	756.0	Anomalies of skull and face bones	1
Herkimer			
	752.6	Hypospadias & epispadias	6
	754.3	Congenital dislocation of hip	5
	745.4	Ventricular septal defect	2
	745.5	Ostium secundum atrial septal defect	2
	747.3	Anomalies of pulmonary artery	2
	754.5	Varus deformities of feet	2
	755.3	Reduction deformities of lower limb	2
	277.0	Cystic fibrosis	1
	740.0	Anencephalus	1
	741.0	Spina bifida with hydrocephalus	1
	742.1	Microcephalus	1
	744.9	Unspecified anomalies of face & neck	1
	745.2	Tetralogy of Fallot	1
	746.3	Congenital stenosis of aortic arch	1
	746.4	Congenital insufficiency of aortic arch	1
	746.8	Other specified anomalies of heart	1
	747.0	Patent ductus arteriosus	1
	748.3	Other anomalies of larynx, trachea, & bronchus	1
	748.5	Agenesis, hypoplasia & dysplasia, lung	1
	749.0	Cleft palate	1
	752.5	Undescended testicle	1
	752.7	Indeterminate sex & pseudo-hermaphroditism	1
	752.8	Other specified anomalies of genital organs	1
	755.2	Reduction deformities of upper limb	1

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County	ICD - 9 Code	Description	Number Reported
Herkimer cont'd			
	755.5	Other anomalies of upper limb including shoulder girdle	1
	755.6	Other anomalies of lower limb including pelvic girdle	1
	756.1	Anomalies of spine	1
	756.7	Anomalies of abdominal wall	1
	757.6	Specified anomalies of breast	1
	759.8	Other specified anomalies	1
Jefferson			
	748.3	Other anomalies of larynx, trachea, & bronchus	6
	750.5	Congenital hypertrophic pyloric stenosis	5
	752.5	Undescended testicle	5
	745.4	Ventricular septal defect	4
	745.5	Ostium secundum atrial septal defect	3
	758.0	Down syndrome	3
	741.0	Spina bifida with hydrocephalus	2
	742.3	Congenital hydrocephalus	2
	742.4	Other specified anomalies of brain	2
	746.8	Other specified anomalies of heart	2
	747.3	Anomalies of pulmonary artery	2
	748.5	Agenesis, hypoplasia & dysplasia, lung	2
	749.2	Cleft palate with cleft lip	2
	753.2	Obstructive defects of renal pelvis & ureter	2
	754.5	Varus deformities of feet	2
Kings			
	745.5	Ostium secundum atrial septal defect	154
	745.4	Ventricular septal defect	117
	752.6	Hypospadias & epispadias	104
	752.5	Undescended testicle	85
	754.5	Varus deformities of feet	76
	754.3	Congenital dislocation of hip	66
	753.2	Obstructive defects of renal pelvis & ureter	61
	746.8	Other specified anomalies of heart	54
	758.0	Down syndrome	45
	746.0	Anomalies of pulmonary valve	44
Lewis			
	752.6	Hypospadias & epispadias	3
	745.2	Tetralogy of Fallot	2
	754.5	Varus deformities of feet	2
	745.4	Ventricular septal defect	1
	745.5	Ostium secundum atrial septal defect	1
	746.8	Other specified anomalies of heart	1
	747.1	Coarctation of aorta	1
	747.3	Anomalies of pulmonary artery	1
	750.5	Congenital hypertrophic pyloric stenosis	1
	753.1	Cystic kidney disease	1
	756.0	Anomalies of skull and face bones	1
Livingston			
	752.5	Undescended testicle	4

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Livingston cont'd			
	745.4	Ventricular septal defect	3
	745.5	Ostium secundum atrial septal defect	2
	746.0	Anomalies of pulmonary valve	2
	749.1	Cleft lip	2
	752.6	Hypospadias & epispadias	2
	753.1	Cystic kidney disease	2
	753.2	Obstructive defects of renal pelvis & ureter	2
	754.3	Congenital dislocation of hip	2
	754.5	Varus deformities of feet	2
Madison			
	754.3	Congenital dislocation of hip	4
	755.6	Other anomalies of lower limb including pelvic girdle	4
	745.5	Ostium secundum atrial septal defect	3
	749.0	Cleft palate	2
	749.2	Cleft palate with cleft lip	2
	750.5	Congenital hypertrophic pyloric stenosis	2
	754.5	Varus deformities of feet	2
	756.0	Anomalies of skull and face bones	2
	190.5	Malignant neoplasm of the retina	1
	343.9	Infantile cerebral palsy unspecified	1
	425.3	Endocardiac fibroelastosis	1
	742.0	Encephalocele	1
	742.1	Microcephalus	1
	742.2	Reduction deformities of brain	1
	742.3	Congenital hydrocephalus	1
	742.4	Other specified anomalies of brain	1
	745.1	Transposition of great vessels	1
	745.3	Common ventricle	1
	745.4	Ventricular septal defect	1
	746.8	Other specified anomalies of heart	1
	747.0	Patent ductus arteriosus	1
	747.3	Anomalies of pulmonary artery	1
	750.3	Tracheoesophageal fistula, esophageal atresia & stenosis	1
	752.5	Undescended testicle	1
	752.6	Hypospadias & epispadias	1
	753.2	Obstructive defects of renal pelvis & ureter	1
	753.5	Exstrophy of urinary bladder	1
	755.1	Syndactyly	1
	755.3	Reduction deformities of lower limb	1
	757.6	Specified anomalies of breast	1
	758.0	Down syndrome	1
	759.8	Other specified anomalies	1
Monroe			
	752.6	Hypospadias & epispadias	39
	745.4	Ventricular septal defect	29
	752.5	Undescended testicle	25
	753.2	Obstructive defects of renal pelvis & ureter	21
	746.8	Other specified anomalies of heart	20
	746.0	Anomalies of pulmonary valve	19
	750.5	Congenital hypertrophic pyloric stenosis	18

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County	ICD - 9 Code	Description	Number Reported
Monroe cont'd			
	754.5	Varus deformities of feet	18
	754.3	Congenital dislocation of hip	15
	745.5	Ostium secundum atrial septal defect	13
Montgomery			
	746.8	Other specified anomalies of heart	4
	745.5	Ostium secundum atrial septal defect	3
	756.0	Anomalies of skull and face bones	3
	746.0	Anomalies of pulmonary valve	2
	748.3	Other anomalies of larynx, trachea, & bronchus	2
	749.0	Cleft palate	2
	750.5	Congenital hypertrophic pyloric stenosis	2
	752.5	Undescended testicle	2
	752.6	Hypospadias & epispadias	2
	742.2	Reduction deformities of brain	1
	742.8	Other specified anomalies of nervous system	1
	745.1	Transposition of great vessels	1
	746.1	Tricuspid atresia & stenosis	1
	746.3	Congenital stenosis of aortic arch	1
	746.5	Congenital mitral stenosis	1
	747.0	Patent ductus arteriosus	1
	747.1	Coarctation of aorta	1
	747.2	Other anomalies of aorta	1
	748.0	Choanal atresia	1
	749.1	Cleft lip	1
	749.2	Cleft palate with cleft lip	1
	751.1	Atresia and stenosis of small intestine	1
	754.3	Congenital dislocation of hip	1
	754.5	Varus deformities of feet	1
	755.3	Reduction deformities of lower limb	1
	759.5	Tuberous sclerosis	1
Nassau			
	753.2	Obstructive defects of renal pelvis & ureter	65
	745.5	Ostium secundum atrial septal defect	62
	745.4	Ventricular septal defect	58
	752.5	Undescended testicle	58
	752.6	Hypospadias & epispadias	58
	750.5	Congenital hypertrophic pyloric stenosis	49
	754.3	Congenital dislocation of hip	36
	754.5	Varus deformities of feet	32
	746.8	Other specified anomalies of heart	25
	746.0	Anomalies of pulmonary valve	20
New York			
	745.4	Ventricular septal defect	86
	754.3	Congenital dislocation of hip	66
	745.5	Ostium secundum atrial septal defect	64
	752.5	Undescended testicle	45
	752.6	Hypospadias & epispadias	41
	754.5	Varus deformities of feet	31
	753.2	Obstructive defects of renal pelvis & ureter	30

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County	ICD - 9 Code	Description	Number Reported
New York cont'd			
	746.8	Other specified anomalies of heart	28
	747.0	Patent ductus arteriosus	26
	746.0	Anomalies of pulmonary valve	22
Niagara			
	752.6	Hypospadias & epispadias	10
	747.3	Anomalies of pulmonary artery	9
	750.5	Congenital hypertrophic pyloric stenosis	6
	752.5	Undescended testicle	6
	745.4	Ventricular septal defect	5
	745.5	Ostium secundum atrial septal defect	5
	753.2	Obstructive defects of renal pelvis & ureter	5
	748.3	Other anomalies of larynx, trachea, & bronchus	4
	754.3	Congenital dislocation of hip	4
	758.0	Down syndrome	4
Oneida			
	756.0	Anomalies of skull and face bones	9
	745.4	Ventricular septal defect	7
	752.6	Hypospadias & epispadias	7
	754.5	Varus deformities of feet	7
	745.5	Ostium secundum atrial septal defect	6
	750.5	Congenital hypertrophic pyloric stenosis	6
	752.5	Undescended testicle	5
	755.2	Reduction deformities of upper limb	5
	758.0	Down syndrome	5
	745.2	Tetralogy of Fallot	4
	753.2	Obstructive defects of renal pelvis & ureter	4
	754.3	Congenital dislocation of hip	4
Onondaga			
	754.3	Congenital dislocation of hip	29
	745.4	Ventricular septal defect	27
	752.5	Undescended testicle	20
	752.6	Hypospadias & epispadias	19
	753.2	Obstructive defects of renal pelvis & ureter	16
	754.5	Varus deformities of feet	16
	745.5	Ostium secundum atrial septal defect	15
	750.5	Congenital hypertrophic pyloric stenosis	13
	746.8	Other specified anomalies of heart	9
	742.3	Congenital hydrocephalus	8
Ontario			
	754.3	Congenital dislocation of hip	8
	752.6	Hypospadias & epispadias	6
	754.5	Varus deformities of feet	6
	752.5	Undescended testicle	5
	753.2	Obstructive defects of renal pelvis & ureter	5
	745.4	Ventricular septal defect	4
	755.6	Other anomalies of lower limb including pelvic girdle	3

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Ontario cont'd			
	758.0	Down syndrome	3
	742.3	Congenital hydrocephalus	2
	746.0	Anomalies of pulmonary valve	2
	746.8	Other specified anomalies of heart	2
	750.5	Congenital hypertrophic pyloric stenosis	2
	752.8	Other specified anomalies of genital organs	2
	755.3	Reduction deformities of lower limb	2
	755.5	Other anomalies of upper limb including shoulder girdle	2
Orange			
	752.5	Undescended testicle	17
	752.6	Hypospadias & epispadias	13
	745.4	Ventricular septal defect	12
	750.5	Congenital hypertrophic pyloric stenosis	11
	745.5	Ostium secundum atrial septal defect	8
	754.3	Congenital dislocation of hip	7
	754.5	Varus deformities of feet	7
	756.0	Anomalies of skull and face bones	7
	747.3	Anomalies of pulmonary artery	6
	742.3	Congenital hydrocephalus	5
Orleans			
	752.6	Hypospadias & epispadias	4
	752.5	Undescended testicle	3
	745.4	Ventricular septal defect	2
	755.6	Other anomalies of lower limb including pelvic girdle	2
	756.0	Anomalies of skull and face bones	2
	359.0	Congenital Hereditary Muscular Dystrophy	1
	741.0	Spina bifida with hydrocephalus	1
	742.2	Reduction deformities of brain	1
	742.5	Other specified anomalies of spinal cord	1
	743.0	Anophthalmos	1
	744.0	Anomalies of ear causing impairment of hearing	1
	745.2	Tetralogy of Fallot	1
	746.0	Anomalies of pulmonary valve	1
	746.4	Congenital insufficiency of aortic arch	1
	747.1	Coarctation of aorta	1
	747.3	Anomalies of pulmonary artery	1
	751.3	Hirschprung's disease & other functional disorders of colon	1
	753.2	Obstructive defects of renal pelvis & ureter	1
	754.2	Deformities of spine	1
	754.3	Congenital dislocation of hip	1
	754.5	Varus deformities of feet	1
	755.2	Reduction deformities of upper limb	1
	756.1	Anomalies of spine	1
	756.3	Other anomalies of ribs and sternum	1
	756.7	Anomalies of abdominal wall	1
	758.0	Down syndrome	1
Oswego			
	745.4	Ventricular septal defect	10

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County	ICD - 9 Code	Description	Number Reported
Oswego cont'd			
	752.6	Hypospadias & epispadias	10
	748.3	Other anomalies of larynx, trachea, & bronchus	7
	750.5	Congenital hypertrophic pyloric stenosis	6
	754.3	Congenital dislocation of hip	5
	752.5	Undescended testicle	4
	754.5	Varus deformities of feet	4
	756.0	Anomalies of skull and face bones	4
	746.0	Anomalies of pulmonary valve	3
	746.8	Other specified anomalies of heart	3
	747.0	Patent ductus arteriosus	3
	753.0	Renal agenesis & dysgenesis	3
Otsego			
	752.5	Undescended testicle	4
	752.6	Hypospadias & epispadias	4
	753.2	Obstructive defects of renal pelvis & ureter	4
	749.0	Cleft palate	2
	754.5	Varus deformities of feet	2
	343.9	Infantile cerebral palsy unspecified	1
	359.0	Congenital Hereditary Muscular Dystrophy	1
	745.4	Ventricular septal defect	1
	745.5	Ostium secundum atrial septal defect	1
	750.3	Tracheoesophageal fistula, esophageal atresia & stenosis	1
	751.1	Atresia and stenosis of small intestine	1
	751.2	Atresia and stenosis of large intestine, rectum, & anal canal	1
	753.0	Renal agenesis & dysgenesis	1
	753.1	Cystic kidney disease	1
	753.3	Other specified anomalies of kidney	1
	753.4	Other specified anomalies of ureter	1
	754.3	Congenital dislocation of hip	1
	759.8	Other specified anomalies	1
Putnam			
	754.3	Congenital dislocation of hip	5
	752.6	Hypospadias & epispadias	3
	742.3	Congenital hydrocephalus	2
	745.4	Ventricular septal defect	2
	745.5	Ostium secundum atrial septal defect	2
	747.0	Patent ductus arteriosus	2
	750.5	Congenital hypertrophic pyloric stenosis	2
	755.6	Other anomalies of lower limb including pelvic girdle	2
	282.0	Hereditary spherocytosis	1
	742.4	Other specified anomalies of brain	1
	744.0	Anomalies of ear causing impairment of hearing	1
	753.2	Obstructive defects of renal pelvis & ureter	1
	753.6	Atresia and stenosis of urethra & bladder neck	1
	756.1	Anomalies of spine	1
	756.8	Other specified anomalies of muscle, tendon, fascia, etc.	1
	757.1	Ichthyosis congenita	1
	757.6	Specified anomalies of breast	1
	758.5	Other conditions due autosomal anomalies	1

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Queens			
	745.5	Ostium secundum atrial septal defect	119
	745.4	Ventricular septal defect	105
	752.6	Hypospadias & epispadias	103
	752.5	Undescended testicle	98
	753.2	Obstructive defects of renal pelvis & ureter	67
	754.3	Congenital dislocation of hip	62
	754.5	Varus deformities of feet	51
	746.8	Other specified anomalies of heart	44
	750.5	Congenital hypertrophic pyloric stenosis	43
	756.0	Anomalies of skull and face bones	30
Rensselaer			
	752.6	Hypospadias & epispadias	13
	752.5	Undescended testicle	5
	754.5	Varus deformities of feet	5
	745.4	Ventricular septal defect	4
	745.5	Ostium secundum atrial septal defect	4
	753.2	Obstructive defects of renal pelvis & ureter	4
	754.3	Congenital dislocation of hip	4
	742.3	Congenital hydrocephalus	3
	746.0	Anomalies of pulmonary valve	3
	748.3	Other anomalies of larynx, trachea, & bronchus	3
	750.5	Congenital hypertrophic pyloric stenosis	3
	752.8	Other specified anomalies of genital organs	3
Richmond			
	745.4	Ventricular septal defect	23
	752.6	Hypospadias & epispadias	19
	754.5	Varus deformities of feet	19
	745.5	Ostium secundum atrial septal defect	18
	752.5	Undescended testicle	18
	754.3	Congenital dislocation of hip	11
	746.8	Other specified anomalies of heart	9
	753.2	Obstructive defects of renal pelvis & ureter	9
	750.5	Congenital hypertrophic pyloric stenosis	8
	756.0	Anomalies of skull and face bones	5
Rockland			
	752.5	Undescended testicle	9
	752.6	Hypospadias & epispadias	9
	745.5	Ostium secundum atrial septal defect	8
	745.4	Ventricular septal defect	7
	750.5	Congenital hypertrophic pyloric stenosis	5
	753.2	Obstructive defects of renal pelvis & ureter	5
	745.2	Tetralogy of Fallot	4
	751.2	Atresia and stenosis of large intestine, rectum, & anal canal	4
	754.5	Varus deformities of feet	4
	748.3	Other anomalies of larynx, trachea, & bronchus	3
	748.5	Agenesis, hypoplasia & dysplasia, lung	3

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County	ICD - 9 Code	Description	Number Reported
Rockland cont'd			
	755.1	Syndactyly	3
	755.6	Other anomalies of lower limb including pelvic girdle	3
	756.7	Anomalies of abdominal wall	3
	758.0	Down syndrome	3
Saratoga			
	745.4	Ventricular septal defect	12
	745.5	Ostium secundum atrial septal defect	10
	752.5	Undescended testicle	10
	754.3	Congenital dislocation of hip	9
	752.6	Hypospadias & epispadias	8
	753.2	Obstructive defects of renal pelvis & ureter	6
	754.5	Varus deformities of feet	6
	746.8	Other specified anomalies of heart	5
	749.1	Cleft lip	4
	750.5	Congenital hypertrophic pyloric stenosis	4
Schenectady			
	752.6	Hypospadias & epispadias	12
	752.5	Undescended testicle	7
	745.4	Ventricular septal defect	5
	745.5	Ostium secundum atrial septal defect	5
	754.3	Congenital dislocation of hip	5
	747.0	Patent ductus arteriosus	4
	753.2	Obstructive defects of renal pelvis & ureter	4
	754.5	Varus deformities of feet	4
	742.1	Microcephalus	3
	742.4	Other specified anomalies of brain	2
	743.3	Congenital cataract & lens anomalies	2
	746.0	Anomalies of pulmonary valve	2
	746.3	Congenital stenosis of aortic arch	2
	751.2	Atresia and stenosis of large intestine, rectum, & anal canal	2
	755.6	Other anomalies of lower limb including pelvic girdle	2
	756.0	Anomalies of skull and face bones	2
Schoharie			
	742.4	Other specified anomalies of brain	2
	746.8	Other specified anomalies of heart	2
	750.5	Congenital hypertrophic pyloric stenosis	2
	742.1	Microcephalus	1
	748.3	Other anomalies of larynx, trachea, & bronchus	1
	749.2	Cleft palate with cleft lip	1
	752.6	Hypospadias & epispadias	1
Schuyler			
	754.5	Varus deformities of feet	2
	745.4	Ventricular septal defect	1
	745.5	Ostium secundum atrial septal defect	1
	747.3	Anomalies of pulmonary artery	1
	749.1	Cleft lip	1
	749.2	Cleft palate with cleft lip	1

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County	ICD - 9 Code	Description	Number Reported
Schuyler cont'd			
	750.5	Congenital hypertrophic pyloric stenosis	1
	751.2	Atresia and stenosis of large intestine, rectum, & anal canal	1
	752.4	Anomalies of cervix, vagina & external female genitalia	1
	753.6	Atresia and stenosis of urethra & bladder neck	1
	754.3	Congenital dislocation of hip	1
	754.4	Congenital genu recurvatum & bowing of long bones of leg	1
	756.0	Anomalies of skull and face bones	1
	756.7	Anomalies of abdominal wall	1
Seneca			
	741.0	Spina bifida with hydrocephalus	2
	745.4	Ventricular septal defect	2
	746.8	Other specified anomalies of heart	2
	754.3	Congenital dislocation of hip	2
	741.9	Spina bifida w/o hydrocephalus	1
	744.0	Anomalies of ear causing impairment of hearing	1
	745.0	Common truncus	1
	745.5	Ostium secundum atrial septal defect	1
	746.6	Congenital mitral insufficiency	1
	747.2	Other anomalies of aorta	1
	750.2	Other specified anomalies, mouth and pharynx	1
	751.2	Atresia and stenosis of large intestine, rectum, & anal canal	1
	752.6	Hypospadias & epispadias	1
	754.5	Varus deformities of feet	1
	755.1	Syndactyly	1
	756.1	Anomalies of spine	1
	756.5	Osteodystrophies	1
	756.6	Anomalies of diaphragm	1
	756.7	Anomalies of abdominal wall	1
	771.1	Congenital cytomegalovirus infection	1
St. Lawrence			
	758.0	Down syndrome	3
	745.5	Ostium secundum atrial septal defect	2
	745.6	Endocardial cushion defects	2
	752.5	Undescended testicle	2
	752.6	Hypospadias & epispadias	2
	743.4	Coloboma & other anomalies of anterior segment	1
	745.2	Tetralogy of Fallot	1
	745.4	Ventricular septal defect	1
	746.8	Other specified anomalies of heart	1
	749.1	Cleft lip	1
	750.5	Congenital hypertrophic pyloric stenosis	1
	753.1	Cystic kidney disease	1
	753.4	Other specified anomalies of ureter	1
	754.5	Varus deformities of feet	1
	755.3	Reduction deformities of lower limb	1
	756.6	Anomalies of diaphragm	1
	756.7	Anomalies of abdominal wall	1

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County	ICD - 9 Code	Description	Number Reported
Steuben			
	754.5	Varus deformities of feet	5
	745.4	Ventricular septal defect	3
	750.5	Congenital hypertrophic pyloric stenosis	3
	754.3	Congenital dislocation of hip	3
	758.0	Down syndrome	3
	746.0	Anomalies of pulmonary valve	2
	749.0	Cleft palate	2
	749.2	Cleft palate with cleft lip	2
	752.6	Hypospadias & epispadias	2
	243	Congenital hypothyroidism	1
	658.8	Amniotic bands	1
	742.3	Congenital hydrocephalus	1
	743.0	Anophthalmos	1
	743.4	Coloboma & other anomalies of anterior segment	1
	745.1	Transposition of great vessels	1
	745.5	Ostium secundum atrial septal defect	1
	745.6	Endocardial cushion defects	1
	746.2	Ebstein's anomaly	1
	747.1	Coarctation of aorta	1
	747.3	Anomalies of pulmonary artery	1
	748.5	Agenesis, hypoplasia & dysplasia, lung	1
	749.1	Cleft lip	1
	750.3	Tracheoesophageal fistula, esophageal atresia & stenosis	1
	751.1	Atresia and stenosis of small intestine	1
	751.3	Hirschprung's disease & other functional disorders of colon	1
	751.4	Anomalies of intestinal fixation	1
	752.5	Undescended testicle	1
	752.8	Other specified anomalies of genital organs	1
	753.2	Obstructive defects of renal pelvis & ureter	1
	753.3	Other specified anomalies of kidney	1
	755.3	Reduction deformities of lower limb	1
	755.5	Other anomalies of upper limb including shoulder girdle	1
	756.6	Anomalies of diaphragm	1
	757.3	Other specified anomalies of skin	1
Suffolk			
	745.5	Ostium secundum atrial septal defect	71
	752.6	Hypospadias & epispadias	63
	745.4	Ventricular septal defect	53
	753.2	Obstructive defects of renal pelvis & ureter	33
	752.5	Undescended testicle	31
	750.5	Congenital hypertrophic pyloric stenosis	29
	758.0	Down syndrome	26
	754.3	Congenital dislocation of hip	24
	746.8	Other specified anomalies of heart	20
Sullivan			
	754.5	Varus deformities of feet	20
	752.6	Hypospadias & epispadias	3
	745.4	Ventricular septal defect	2
	745.5	Ostium secundum atrial septal defect	2
	750.5	Congenital hypertrophic pyloric stenosis	2

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Ten Most Frequently Reported Major Malformations
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Congenital Malformations Registry

County	ICD - 9 Code	Description	Number Reported
Sullivan cont'd			
	744.0	Anomalies of ear causing impairment of hearing	1
	745.0	Common truncus	1
	745.1	Transposition of great vessels	1
	746.0	Anomalies of pulmonary valve	1
	747.0	Patent ductus arteriosus	1
	748.3	Other anomalies of larynx, trachea, & bronchus	1
	752.5	Undescended testicle	1
	753.0	Renal agenesis & dysgenesis	1
	753.1	Cystic kidney disease	1
	753.2	Obstructive defects of renal pelvis & ureter	1
	753.4	Other specified anomalies of ureter	1
	754.3	Congenital dislocation of hip	1
	754.5	Varus deformities of feet	1
	755.6	Other anomalies of lower limb including pelvic girdle	1
	756.0	Anomalies of skull and face bones	1
Tioga			
	745.4	Ventricular septal defect	5
	745.5	Ostium secundum atrial septal defect	5
	746.3	Congenital stenosis of aortic arch	3
	746.0	Anomalies of pulmonary valve	2
	754.5	Varus deformities of feet	2
	243	Congenital hypothyroidism	1
	277.0	Cystic fibrosis	1
	742.1	Microcephalus	1
	742.3	Congenital hydrocephalus	1
	745.1	Transposition of great vessels	1
	745.6	Endocardial cushion defects	1
	746.4	Congenital insufficiency of aortic arch	1
	746.8	Other specified anomalies of heart	1
	747.0	Patent ductus arteriosus	1
	747.1	Coarctation of aorta	1
	747.2	Other anomalies of aorta	1
	752.6	Hypospadias & epispadias	1
	754.3	Congenital dislocation of hip	1
	755.5	Other anomalies of upper limb including shoulder girdle	1
	756.0	Anomalies of skull and face bones	1
	758.0	Down syndrome	1
Tompkins			
	745.4	Ventricular septal defect	5
	752.5	Undescended testicle	3
	753.2	Obstructive defects of renal pelvis & ureter	3
	750.5	Congenital hypertrophic pyloric stenosis	2
	751.2	Atresia and stenosis of large intestine, rectum, & anal canal	2
	752.6	Hypospadias & epispadias	2
	754.5	Varus deformities of feet	2
	343.9	Infantile cerebral palsy unspecified	1
	742.3	Congenital hydrocephalus	1
	742.4	Other specified anomalies of brain	1
	746.0	Anomalies of pulmonary valve	1

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County	ICD - 9 Code	Description	Number Reported
Tompkins cont'd			
	746.8	Other specified anomalies of heart	1
	748.0	Choanal atresia	1
	749.1	Cleft lip	1
	751.1	Atresia and stenosis of small intestine	1
	753.4	Other specified anomalies of ureter	1
	756.0	Anomalies of skull and face bones	1
	756.3	Other anomalies of ribs and sternum	1
Ulster			
	752.5	Undescended testicle	7
	752.6	Hypospadias & epispadias	6
	745.4	Ventricular septal defect	4
	745.5	Ostium secundum atrial septal defect	4
	753.2	Obstructive defects of renal pelvis & ureter	4
	754.3	Congenital dislocation of hip	4
	755.6	Other anomalies of lower limb including pelvic girdle	4
	745.2	Tetralogy of Fallot	3
	746.8	Other specified anomalies of heart	3
	747.3	Anomalies of pulmonary artery	3
	754.5	Varus deformities of feet	3
Warren			
	746.8	Other specified anomalies of heart	5
	754.5	Varus deformities of feet	5
	745.5	Ostium secundum atrial septal defect	4
	752.6	Hypospadias & epispadias	4
	745.4	Ventricular septal defect	3
	753.2	Obstructive defects of renal pelvis & ureter	3
	754.3	Congenital dislocation of hip	3
	756.7	Anomalies of abdominal wall	3
	759.8	Other specified anomalies	3
	742.5	Other specified anomalies of spinal cord	2
	746.0	Anomalies of pulmonary valve	2
Washington			
	745.4	Ventricular septal defect	2
	752.6	Hypospadias & epispadias	2
	742.3	Congenital hydrocephalus	1
	742.4	Other specified anomalies of brain	1
	743.4	Coloboma & other anomalies of anterior segment	1
	745.2	Tetralogy of Fallot	1
	746.8	Other specified anomalies of heart	1
	748.5	Agenesis, hypoplasia & dysplasia, lung	1
	750.5	Congenital hypertrophic pyloric stenosis	1
	753.2	Obstructive defects of renal pelvis & ureter	1
	753.4	Other specified anomalies of ureter	1
	754.3	Congenital dislocation of hip	1
	756.6	Anomalies of diaphragm	1
	756.7	Anomalies of abdominal wall	1
Wayne			
	752.5	Undescended testicle	7

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County	ICD - 9 Code	Description	Number Reported
Wayne cont'd			
	745.4	Ventricular septal defect	6
	750.5	Congenital hypertrophic pyloric stenosis	5
	758.0	Down syndrome	5
	752.6	Hypospadias & epispadias	3
	753.2	Obstructive defects of renal pelvis & ureter	3
	745.1	Transposition of great vessels	2
	747.0	Patent ductus arteriosus	2
	754.3	Congenital dislocation of hip	2
	742.1	Microcephalus	1
	742.2	Reduction deformities of brain	1
	743.4	Coloboma & other anomalies of anterior segment	1
	745.2	Tetralogy of Fallot	1
	745.6	Endocardial cushion defects	1
	746.0	Anomalies of pulmonary valve	1
	746.1	Tricuspid atresia & stenosis	1
	746.4	Congenital insufficiency of aortic arch	1
	746.8	Other specified anomalies of heart	1
	747.3	Anomalies of pulmonary artery	1
	748.5	Agenesis, hypoplasia & dysplasia, lung	1
	749.0	Cleft palate	1
	750.3	Tracheoesophageal fistula, esophageal atresia & stenosis	1
	751.1	Atresia and stenosis of small intestine	1
	751.2	Atresia and stenosis of large intestine, rectum, & anal canal	1
	751.4	Anomalies of intestinal fixation	1
	754.5	Varus deformities of feet	1
	757.6	Specified anomalies of breast	1
	771.2	Other congenital infections	1
Westchester			
	752.5	Undescended testicle	39
	752.6	Hypospadias & epispadias	36
	745.4	Ventricular septal defect	31
	745.5	Ostium secundum atrial septal defect	31
	754.3	Congenital dislocation of hip	27
	750.5	Congenital hypertrophic pyloric stenosis	19
	753.2	Obstructive defects of renal pelvis & ureter	19
	747.0	Patent ductus arteriosus	13
	754.5	Varus deformities of feet	13
	742.3	Congenital hydrocephalus	12
Wyoming			
	752.6	Hypospadias & epispadias	4
	755.6	Other anomalies of lower limb including pelvic girdle	4
	752.5	Undescended testicle	3
	745.4	Ventricular septal defect	2
	754.3	Congenital dislocation of hip	2
	343.9	Infantile cerebral palsy unspecified	1
	742.4	Other specified anomalies of brain	1
	743.3	Congenital cataract & lens anomalies	1
	743.5	Congenital anomalies of posterior segment of eye	1
	743.8	Other specified anomalies of eye	1

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County	ICD - 9 Code	Description	Number Reported
Wyoming cont'd			
	744.0	Anomalies of ear causing impairment of hearing	1
	744.9	Unspecified anomalies of face & neck	1
	745.5	Ostium secundum atrial septal defect	1
	745.6	Endocardial cushion defects	1
	746.8	Other specified anomalies of heart	1
	747.3	Anomalies of pulmonary artery	1
	749.0	Cleft palate	1
	749.2	Cleft palate with cleft lip	1
	750.5	Congenital hypertrophic pyloric stenosis	1
	751.2	Atresia and stenosis of large intestine, rectum, & anal canal	1
	752.0	Anomalies of ovaries	1
	752.8	Other specified anomalies of genital organs	1
	753.2	Obstructive defects of renal pelvis & ureter	1
	753.3	Other specified anomalies of kidney	1
	753.7	Anomalies of urachus	1
	754.5	Varus deformities of feet	1
	756.0	Anomalies of skull and face bones	1
	756.7	Anomalies of abdominal wall	1
	758.0	Down syndrome	1
	759.8	Other specified anomalies	1
Yates			
	754.3	Congenital dislocation of hip	2
	743.1	Microphthalmos	1
	745.4	Ventricular septal defect	1
	747.1	Coarctation of aorta	1
	752.6	Hypospadias & epispadias	1
	758.0	Down syndrome	1
	771.2	Other congenital infections	1

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.

The most striking difference among the registries is the low prevalence of anencephaly in New York State. This probably is largely due to the inclusion of stillborn infants in the MACDP and the CBDMP. The CMR includes only live born children (see Section VI, Current Topics). Underreporting is not the only reason for possible differences. The prevalences of some birth defects differ by race. True geographic differences may also exist[3].

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

MACDP Code	Malformation	CMR 1994	MACDP 1985-1993	CBDMP 1983-1989
A01	Anencephalus	0.4	2.5	3.0
A04	Spina bifida	3.6	4.9	5.9
A15	Hydrocephalus	7.7	9.7	5.6
A13	Encephalocele	0.7	1.5	1.5
A16	Microcephalus	5.5	5.5	1.2
B01	An/Microphthalmos	1.1	3.6	3.6
D01	Common truncus	0.8	0.7	1.3
D02	Trans of great vessels	4.1	4.3	5.3
D03	Tetralogy of Fallot	4.7	3.9	3.6
D05	Ventricular septal defect	29.4	29.7	--
D26	Coarctation of aorta	2.6	4.3	--
E06	Lung agenesis/hypoplasia	3.2	5.9	5.7
F01	Cleft palate	6.3	5.3	7.7
F02	Cleft lip ± cleft palate	9.0	8.9	11.0
E01	Choanal atresia	1.5	1.4	1.6
F09	Esophageal/tracheoesophageal atresia fistula	2.6	2.4	2.7
F16	Rectal/large intestine atresia	2.8	4.1	4.6
F08	Pyloric stenosis	14.2	15.3	20.2
F17	Hirschsprung's disease	1.6	2.0	1.4
F21	Biliary atresia	0.4	0.7	0.8
H01	Renal agenesis/hypoplasia	2.8	3.7	4.1
H08	Bladder exstrophy	0.3	0.3	--
H06	Obstructed renal pelvis & ureter	18.3	10.8	7.1
G02	Hypo/epispadias	31.4	37.0	--
K01	Reduct deform of upper limb	3.3	3.3	--
K02	Reduct deform of lower limb	2.8	1.6	--
J11	Arthrogryposis multiplex cong.	1.5	--	1.9
N04	Gastroschisis	1.3	2.5	1.7
N02	Omphalocele	1.3	2.5	2.7
N01	Diaphragmatic hernia	2.1	2.7	3.1
R01	Down syndrome	8.7	9.9	9.6
R02	Trisomy 13	0.8	1.2	1.0
R03	Trisomy 18	1.0	1.9	1.6
S02	Fetal alcohol syndrome	2.0	2.3	1.5
K05	Amniotic bands	0.3	2.0	--

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

3. 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. *MMWR 1993*; 42(No. SS-1):1-7.

Section VI
Current Topics

Follow-up of Children with Birth Defects

Healthy People 2000[1] has three major areas of health promotion and disease prevention objectives: (1) increase the span of healthy life for Americans; (2) reduce the health disparity among Americans; and (3) achieve access to preventive services for all Americans. A special category has been established for surveillance and data systems to help track the nation's progress toward these goals. Good data are needed to monitor progress, especially for vulnerable populations such as people with low income, minority groups and people with certain disabilities. Birth defects registries can help to provide needed data on the vulnerable population of children with congenital abnormalities. While primary prevention is sometimes possible (e.g., folic acid to prevent neural tube defects), for the near future, secondary and tertiary prevention methods will continue to be important to reduce the burden of birth defects on the individual and society.

Mortality in Children with Birth Defects

Nationwide, birth defects are the leading cause of infant mortality and this has been true for more than two decades[2,3]. More than 70% of infants who die, die in the first month of life. The mortality due to birth defects has declined more slowly then has mortality due to preterm birth or low birthweight.

Data on infant mortality in children with major malformations usually come from vital records[3,4]. Birth defects registries can offer improvements over vital records when examining infant mortality in children with birth defects. Vital records are limited by a lack of completeness and accuracy in the reporting of birth defects. For specific malformations, some studies use a case series from a hospital. However, these sources may be biased in their ascertainment. Unless all cases were born in the study hospital, mortality would be underestimated[5]. A hospital case series would miss those cases born at outside institutions, considered unlikely to survive and never transferred for treatment.

The CMR examined the infant mortality experience of infants with birth defects born from 1983 to 1988 in New York State[5]. Infants with major malformations with a mortality of 68 per 1,000 live births had a risk of death 6.2 times higher than the general population risk of 11. Among children with major malformations, mortality risk was increased if the malformation affected chromosome, respiratory system or central nervous system; if the child had more than one major malformation; or if the child was low birthweight. Race did not show the traditional 2:1 black white ratio seen for general infant mortality. After adjusting for birthweight, the risk was the same for white and black infants with major malformations. When cause of death was examined, children with major malformations had higher mortality due to causes other than congenital anomalies. Low birthweight, respiratory distress syndrome, other perinatal causes, infectious diseases and other system causes were all higher in children with major malformations. Only external causes (injuries) and sudden infant death syndrome were not higher in children with major malformations.

Mortality will vary greatly by the type of malformation as well as the number of malformations. The CMR also examined the mortality experience of children with oral clefts, i.e., cleft palate and cleft lip with or without cleft palate[6]. Infants with oral clefts born from 1983 to 1990 to New York State residents were compared with infants without major malformations. The mortality in the first year of life for children with oral clefts and no other major malformations was similar to that for children with no major malformations. There also was no excess mortality up to age five years. However, 35% of the children with oral clefts had other birth defects and experienced mortality of about 20 times higher than for children without major malformations.

Table 1 shows the first year of life mortality for children with specific malformations in the 1994 birth cohort with the mortality for the 1984 cohort used as a comparison. Infant mortality for the general population declined by 30% from 1984 to 1994. Changes in mortality varied considerably by malformation. Spina bifida, diaphragmatic hernia and atresia of the small intestine declined by more than 50%. However, atresia of the large intestine and Teratology of Fallot showed no improvement. These numbers do not take into account whether other malformations were present, which greatly

affects mortality. A condition such as atresia of the large intestine includes rectal atresia that is frequently associated with other malformations. These numbers also reflect only the mortality experience of live born infants. The mortality of conditions such as spina bifida and diaphragmatic hernia may be affected by terminations. It is possible that new technologies such as extra corporeal membrane oxygenation may be involved in the improved survival of infants with diaphragmatic hernia. A series of studies to further examine mortality in children with various malformations is planned.

Tracking Children with Birth Defects

As the CMR was established in the wake of the Love Canal investigation, it is located in the Center for Environmental Health section of the Department of Health. However, birth defect registries can provide useful information to Maternal and Child Health programs as well. Tracking affected children to ensure quality care and enrollment in intervention programs is an important part of disability prevention. To ensure that children with malformations have the best chance to fulfill their potential, they need early access to good medical care and other services such as early intervention. New York State has programs to identify children who may need early intervention. The CMR matched children reported to the CMR to enrollment in early intervention programs to see if these children are receiving services. Table 2 shows the percentage of children with various birth defects who are enrolled in early intervention programs. The CMR is working with the Maternal and Child Health programs in the Department of Health to develop ways to track children in the CMR and insure that their families know about available services.

Section VI - Table 1

First Year of Life Mortality* for Children with Selected Malformations
New York State Congenital Malformations Registry, 1984 and 1994

MALFORMATION	1984	1994	% Change
Spina Bifida	18.6	8.7	-53
Transposition of Great Vessels	27	23.5	-13
Teratology of Fallot	15.7	16.9	8
Hypoplastic Left Heart	88	60.5	-31
Total Anomalous Pulmonary Venous Return	34.8	31.2	-10
Tracheo/esophageal fistula/atresia	19.7	12.3	-38
Diaphragmatic Hernia	55.2	26.5	-52
Atresia of small intestine	17.6	7.7	-56
Atresia of large intestine	13.6	13.2	-3
Total Livebirths	1	0.7	-30

* per 100 livebirths

Section VI - Table 2

Children with Selected Malformations Matched to Early Intervention Program 1994

Malformation	**Percentage of Children in Early Intervention Program**
Spina Bifida	48.9
Microcephalus	41.3
Hydrocephalus	56.1
Ear Anomalies With Impairment of Hearing	40
Transposition of Great Vessels	23.1
Teratology of Fallot	36.4
Cleft Lip and Palate	30.8
Scoliosis	36.8
Hip Dislocation - Combine	5.9
Talipes Equinovarus	26.7
Limb Reduction - Upper	45.9
Limb Reduction - Lower	29.1
Arthrogyposis	45.7
Down Syndrome	77.8
Syndromes	56.1
Fetal Alcohol Syndrome	25

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5. Druschel CM. A descriptive study of prune belly in New York State, 1983 to 1989. *Arch Pediatr Adolesc Med* 1995; 149:70-76.

6. Druschel C, Hughes JP, Olsen C. Mortality among infants with congenital malformations, 1983-1988. *Public Health Reports* 1996; 111:359-365.

7. Druschel CM, Hughes JP, Olsen CL. First year of life mortality among infants with oral clefts; New York State; 1983-1990. *Cleft Palate Cranio-facial Journal*; 1996; 33:400-405.

Abstracts of CMR Recent Publications

1. Applegate MS, Druschel CM. The epidemiology of infantile hypertrophic pyloric stenosis in New York State: 1983-1990. *Arch Pediatric Adolesc Med* 1995; 149:1123-1129.

Objective. To investigate an apparent decline in infantile hypertrophic pyloric stenosis (IHPS) and to examine the characteristics of children with IHPS and any associated malformations.
Design. Cohort study, children with IHPS are compared with the population of live births. Trends of IHPS compared in two data sets: a population-based birth defects registry and hospital discharge data.
Participants. Children with IHPS identified from a birth defects registry and the population of infants live born to residents of New York State from 1983 to 1990.
Main Outcome Measure. Trends in the incidence of IHPS in two data sets, demographic characteristics and malformations associated with IHPS.
Results. IHPS declined from 2.4 per 1,000 live births in 1984 to 1.7 in 1990. White race and male gender were associated with a higher occurrence of IHPS; high birth order, older maternal age, higher maternal education and low birthweight were associated with lower occurrence. Seven percent of children with IHPS had a major malformation compared with 3.7% of the general population. Three major malformations occurred more frequently in children with IHPS: intestinal malrotation, obstructive defects of the urinary tract and esophageal atresia. Fewer cases were found in the birth defects registry compared with hospital discharge data.
Conclusions. There is underreporting of IHPS to the birth defects registry accounting for some of the decline. Children with IHPS have more major malformations compared with the general population, although some of the excess could be attributed to increased detection. Further investigation is needed into the environmental and socioeconomic factors associated with IHPS.

2. Druschel CM. A descriptive study of prune belly in New York State, 1983-1989. *Arch Pediatr Adolesc Med* 1995; 149:70-76.

Objective. To determine the prevalence and spectrum of prune belly in a defined population.
Design. Population-based descriptive study using New York State's Congenital Malformations Registry (CMR).
Setting. The CMR is a statewide registry of children diagnosed with congenital anomalies before the age of two years. Cases were children with prune belly born during the years 1983 to 1989 to women resident in New York State and verified by medical record review.
Main Outcome Measures. The live birth prevalence of prune belly for the total population and for population subgroups, such as race, sex, plurality, maternal age. The occurrence of other malformations with prune belly.
Results. Sixty cases were ascertained, 50 male and 10 female. The live birth prevalence was 3.2 per 100,000 and declined over the time period. The prevalence was higher in males, 5.1 per 100,000 than females, 1.1; and higher in blacks, 5.8 than whites, 2.6. The live birth prevalence in twins, 12.2 per 100,000, was four times higher than in singletons, 3.0. More than 60% of cases died, most in the first week. Seventy percent of cases had one of the commonly described associated defects. Pulmonary hypoplasia was the most common. Almost one-third of the cases had defects other than those typically associated with prune belly.
Conclusions. Twins, black and younger mothers appear to be at higher risk. Mortality remains high, especially early, with many deaths due to pulmonary hypoplasia. Further studies should include stillborn and terminated cases.

3. Druschel CM, Hughes J, Olsen CL. Mortality among infants with congenital malformations in New York State, 1983-1988. *Public Health Rep* 1996; 111:359-365.

While the majority of infant deaths are due to congenital anomalies, few studies specifically look at their mortality experience. This study examines the mortality of infants born from 1983 to 1988 ascertained from a statewide population-based Congenital Malformations Registry and risk factors for first-year mortality. Variables analyzed were the year of birth, birthweight, gestational age, infant sex, number of malformations, number of organ systems, level of care of the birth hospital, maternal age, maternal race and maternal education. Infants with major malformations had a risk of death 6.3 times higher than the general population of live births. The risk declined from 6.4 in 1983 to 5.9 in 1988. The survival for whites and blacks after adjusting for other factors was similar. Having a malformation outweighs most of the other usual risks for infant mortality.

4. Druschel CM, Hughes JP, Olsen CL. First year of life mortality among infants with oral clefts: New York State, 1983-1990. *Cleft Palate Craniofacial J* 1996; 33(5):400-405.

This study examined the mortality experience of children with oral clefts using the New York State Congenital Malformations Registry. Infants born in the years 1983 to 1990 to New York residents, diagnosed with an oral cleft and matched to their birth certificate were included in the analysis. Children with oral clefts were compared with a sample of live births from the years 1983-1990 without malformation. Children with cleft palate without additional malformations had a statistically nonsignificant adjusted risk of 1.2 when compared with children with no malformations. Children with cleft lip with or without cleft palate had a 1.1 adjusted risk. However, 35% of children with oral clefts had associated malformations and experienced much higher mortality. Children with oral clefts should be carefully evaluated for additional malformations. If none are found, their mortality appears not to be elevated.

5. Olsen CL, Polan AK, Cross PK. Case ascertainment for state-based birth defects registries: characteristics of unreported infants ascertained through birth certificates and their impact on registry statistics in New York State. *Paediatr Perinat Epidemiol* 1996; 10:161-174.

Summary. Cases in the New York State Congenital Malformations Registry are reported by hospitals and physicians. This study was undertaken to determine whether case finding should be expanded to include routine matching of vital records files to the registry to identify unreported children. Matching of children who were born in 1983-1986 and who had a congenital malformation noted on their birth certificate yielded 2,837 children who were not in the registry. The hospital of record was asked to submit a registry report if the child's medical record contained a congenital malformation. Medical records for 1,267 (45%) of these children indicated that the child was normal, with no mention of a malformation. Medical records could not be located for 137. Registry reports were submitted for 1,433; 67 of whom were subsequently found in the registry, leaving 1,366 bona fide new cases. These new cases differ significantly from registry cases for a number of birth certificate variables and type of congenital malformation. The birth certificate cases were more likely than registry cases to have only one malformation and to have only a minor malformation. The 1,366 new cases comprised 2.1% of all registry cases for 1983-1986. Their addition increased the statewide prevalence of major malformations by 1.7%, from 416.5 to 423.4 per 10,000 live births. Except for anencephaly, the prevalence of specific malformations was not altered measurably by the addition of these cases. Lengthy and continuous follow-up was required to obtain registry reports. The small number of cases found does not seem to justify the amount of resources that would be required to routinely use birth certificates to augment case finding in New York State.

6. Olsen CL, Cross PK, Gensburg LJ, Hughes JP. The Effects of Prenatal Diagnosis, Population Aging, and Changing Fertility Rates on the Live Birth Prevalence of Down Syndrome in New York State, 1983-1992. *Prenatal Diagnosis* 1996; 16:991-1002.

Abstract. The incidence of Down syndrome (DS) at conception is highly dependent upon the maternal age distribution and age-specific pregnancy rates. Live birth prevalence of DS reflects these factors and fetal deaths. This study examined DS live birth prevalence from 1983-1992 in New York State and analyzed the effects of demographic changes and prenatal diagnosis use on the observed live birth prevalence. Expected DS live birth prevalence without prenatal diagnosis was calculated and compared to the observed prevalence. Data were obtained from the birth defects registry, vital records and population data maintained by the New York State Department of Health. Over time, DS live birth prevalence was stable at about 10.4 per 10,000 live births. The percentage and number of women in the population above age 30 increased, as did birth rates among these women. Birth rates among younger women decreased. The proportion of DS babies born to women aged 35 and over increased from 27.1% to 34.1%. Use of prenatal diagnosis by this age group ranged from 39.6% to 43.2%, and increased steadily from 1.8% to 4.3% among women under 35. Detection of DS fetuses increased from 82 in 1985 to 233 in 1992. Without prenatal diagnosis, DS live birth prevalence in 1992 would have reached 15.3 per 10,000 live births compared with the 10.2 observed. Prenatal diagnosis has prevented an increase in DS live birth prevalence but has not been sufficient to reduce live birth prevalence significantly.

7. Olsen C. L., Hughes, J. P., Youngblood, L. G., and Sharpe-Stimac, M. Epidemiology of Holoprosencephaly and Phenotypic Characteristics of Affected Children: New York State, 1984-1989. *American Journal of Medical Genetics* 1997; 73:217-226.

Abstract. Holoprosencephaly is a congenital defect of the median structures of the brain and face. The epidemiology is poorly known due to the paucity of population-based studies. This study describes the epidemiology of holoprosencephaly in a large population, using cases identified through the New York State Congenital Malformations Registry. The authors describe the range and co-occurrence of craniofacial abnormalities present, and examine the correspondence between the severity of craniofacial abnormalities, chromosomal abnormalities and severity of the brain defect. Liveborn cases totaled 78, yielding a prevalence of 4.8 per 100,000 live births. Prevalence among females was nearly double that in males, and was 4.2 times higher among infants of mothers under age 18 compared with infants of older mothers. Only 9.8% of all cases had no craniofacial abnormalities other than the brain defect. Eye malformations were present in 76.8%, nose malformations in 69.5%, ear malformations in 50% and oral clefts in 41.5%. These malformations arise at different times during gestation. The variability in patterns of their co-occurrence suggests variability in the time during gestation when holoprosencephaly arises, which in turn supports a model of multiple causality. Children with alobar holoprosencephaly tended to have the most severe craniofacial anomalies, but the correspondence was not 100%. Similarly, craniofacial phenotype does not discriminate between cytogenetically normal and abnormal cases.

APPENDICES

Appendix 1

Reporting Card, Congenital Malformations Registry

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. Minor anomalies are 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.

This 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
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 birthweight 2500 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, < 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

Minor Malformations, continued.

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

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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 that 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.

Table 1

Birth Certificate Matching by Place of Birth
1994 Births

Region	Matched	Not Matched
Outside New York City	95.7	4.3
New York City	92.8	7.2
New York State	94.3	5.7

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 (used in Section V) 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