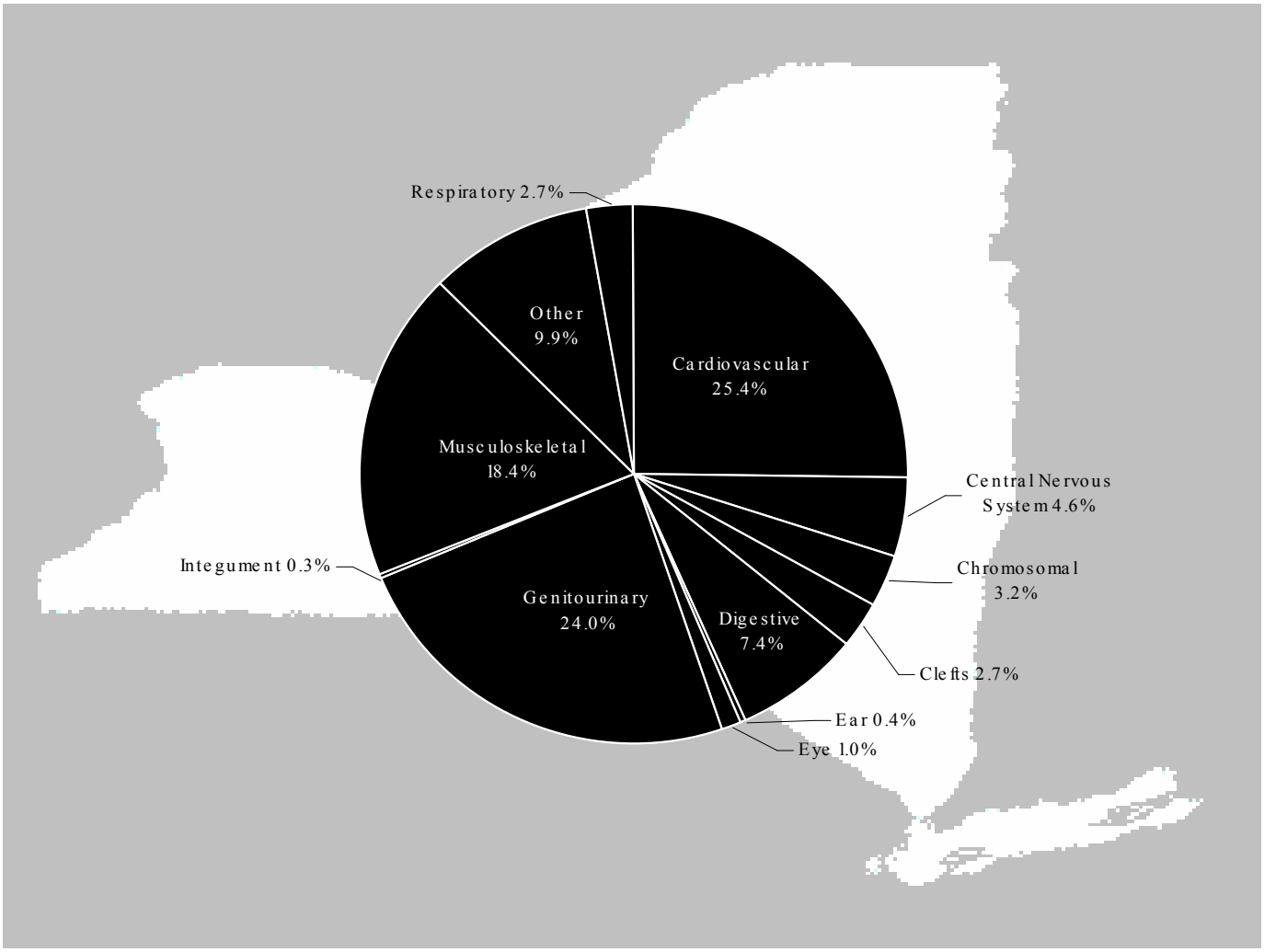
New York State Department of Health

Congenital Malformations Registry Summary Report



Statistical Summary of Children Born in 1998 - 2001 and Diagnosed Through 2003

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

Comments regarding the format or content of this report are welcome.

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New York State Department of Health

Congenital Malformations Registry

Summary Report

Statistical Summary of Children Born in 1998-2001 and Diagnosed Through 2003

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Summary

This Congenital Malformations Registry Summary Report presents rates of congenital malformations occurring among the 1,024,714 children who were born alive to New York residents in 1998-2001. The children reported with a major congenital malformation represent 4.1% of live births. Males had a higher rate of major congenital malformations than females (5.0% versus 3.2%), and black children had a higher major malformation rate than white children (4.8% versus 3.8%). This information is provided through mandated reporting by hospitals and physicians.

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

This is the fifteenth report from the CMR. Reports are also available by request for the 1983 to 1997 birth cohorts. This report and the reports for 1994-1997 are also available on the Department of Health website. The statistics in this report are **not** comparable to reports before 1992. In 1992, the CMR began to use a new coding system that allows for greater detail in coding. For previous years, ICD-9 codes were used. Information from birth certificates was used to supplement or correct reported data. Birth certificate matching also helps eliminate duplicate cases reported under different names and nonresident births. Reports produced for 1989 to 1991 did not use birth certificate matching.

PROGRAM OVERVIEW

Background

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

Although radiation and rubella had been linked to birth defects, not until the thalidomide tragedy of the early 1960s was there a widespread interest in possible associations between congenital malformations and environmental agents. During the 1970s, interest continued to grow in birth defects and birth defects surveillance as a result of the growing recognition of the problems of toxic waste dumps such as Love Canal and accidents such as Three Mile Island and Seveso. In response, many states began to develop birth defects registries in order to have data for tracking trends in malformation rates.[6,7] A birth defects registry also makes it possible to respond to public concerns about possible excess occurrence of malformations with timely, objective investigations. A birth defects registry can provide cases for traditional epidemiologic studies of specific 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 in 1978 to 1983 confirmed the inadequacies of relying on birth certificates to monitor and evaluate birth defects.

New York's Congenital Malformations Registry was established by enactment of Part 22 of the State Sanitary Code in 1981. Reporting to the registry began in October 1982. Hospitals and physicians are required to report children under two years of age diagnosed with a malformation. The majority of reports are sent by hospitals, primarily from their medical records departments. A small number are sent by individual physicians to verify 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 (see Appendix 1) and electronically on the Internet using the Health Provider Network (HPN). The Department of Health developed the HPN as a secure system for electronically collecting and distributing health related data. 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 along with electronic reports to a DOH UNIX server for updating of the CMR database.

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.

1998-2001 Report

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

The statistics in this report are **not** comparable to reports prior to 1992. The 1998-2001 report is based on birth certificate matched cases with resident live births from the vital records file used as the denominator. The available birth certificate fields are used to supplement or correct reported data. Birth certificate data are used to establish maternal residence at birth. Birth certificate matching helps eliminate duplicate cases reported under different names. Racial data are not comparable because race is defined by maternal race from the birth certificate. Using maternal race is a common practice among birth defects registries nationwide as the race of the father is poorly reported. In earlier years, race was defined by what was reported on the CMR form, which may differ from what is recorded on the birth certificate. In 1992, the registry began using a new coding system, the modified British Pediatric Association code (BPA). This coding scheme is used by a number of other congenital malformations registries and allows for greater specificity than does the ICD-9 system. Since 1992, the list of major malformations has been revised (see Appendix 4) changing the list of major malformations used in Sections I and II and the number of specific malformation prevalences in Section III.

CMR Birth Cohort reports are intended as a resource for programs providing primary, secondary and tertiary preventive health care and for public officials concerned with reducing overall mortality and morbidity. The first annual cohort included children born in 1983 and reported with a malformation diagnosed before their second birthday.[9] This report describes children born in 1998-2001 and diagnosed before their second birthday. Reports are also available for the 1984 through 1997 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 the U.S. 2000 census, the population of New York State was about 19.0 million; more than 42% 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 1998-2001, there were 1,024,714 resident live births reported to the state's vital registration, 20.8% to black mothers, and 20.8% to Hispanic mothers. In accordance with the practices of other state birth defects registries, the race of the child is based on race of the mother only. Nearly 37.9% of live births were to New York City residents.

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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 1998 to 2001 and reported to the registry with major malformations. Since a new coding system began to be used in 1992, the list of major malformations has been revised (see Appendix 4). Thus, the prevalence in this report are not comparable to reports prior to 1992.

The overall occurrences of major malformations for the four years ranged from 4.1% to 4.2% of live births. Male children have a higher rate of major malformations than female children (4.8% to 5.0% versus 3.1% to 3.4%, Tables 1a, 1b, 1c, 1d). This difference is consistent within different racial groups. The rates for major malformations are somewhat higher for black than for white children (4.8% to 4.9% versus 3.9% to 4.2%). The major malformation rate among children with residence at birth in New York State excluding New York City was slightly higher than children with residence at birth in New York City (4.1% to 4.3% versus 3.8% to 4.1%). The smaller number of births in the "other" racial category makes these rates difficult to interpret.

About 80% of children reported with major malformations have only one major malformation (Tables 2a, 2b, 2c, 2d). Since most children had one major malformation, the race-sex patterns seen for all major malformations are similar to the patterns seen in children with a single major malformation (Tables 3a, 3b, 3c, 3d). All race-sex groups for children with multiple major malformations showed little variation (Tables 4a, 4b, 4c, 4d)

Section 1 - Table 1a
1998 Births - New York State Residents
Percent of Live Births with One or More Major Malformations
Sex by Race and Residence

	Both Sexes			Males			Females		
Race and Residence	Infants	Total Births	%	Infants	Total Births	%	Infants	Total Births	%
New York State									
- All Races	10,902	257,737	4.2	6,638	132,260	5.0	4,264	125,477	3.4
- White	7,670	184,517	4.2	4,771	94,904	5.0	2,899	89,613	3.2
- Black	2,645	54,108	4.9	1,503	27,523	5.5	1,142	26,585	4.3
- Other	551	17,930	3.1	342	9,208	3.7	209	8,722	2.4
- Unknown/Missing	36	1,182	3.0	22	625	3.5	14	557	2.5
NYS Excluding NYC									
- All Races	6,881	160,645	4.3	4,263	82,403	5.2	2,618	78,242	3.3
- White	5,391	129,487	4.2	3,396	66,535	5.1	1,995	62,952	3.2
- Black	1,284	25,012	5.1	744	12,766	5.8	540	12,246	4.4
- Other	183	5,357	3.4	106	2,683	4.0	77	2,674	2.9
- Unknown/Missing	23	789	2.9	17	419	4.1	6	370	1.6
New York City									
- All Races	4,021	97,092	4.1	2,375	49,857	4.8	1,646	47,235	3.5
- White	2,279	55,030	4.1	1,375	28,369	4.8	904	26,661	3.4
- Black	1,361	29,096	4.7	759	14,757	5.1	602	14,339	4.2
- Other	368	12,573	2.9	236	6,525	3.6	132	6,048	2.2
- Unknown/Missing	13	393	3.3	5	206	2.4	8	187	4.3

Section 1 - Table 1b
1999 Births - New York State Residents
Percent of Live Births with One or More Major Malformations
Sex by Race and Residence

	Both Sexes			Males			Females		
Race and Residence	Infants	Total Births	%	Infants	Total Births	%	Infants	Total Births	%
New York State									
- All Races	10,342	255,147	4.1	6,340	130,758	4.8	4,002	124,389	3.2
- White	7,147	182,448	3.9	4,500	93,870	4.8	2,647	88,578	3.0
- Black	2,529	53,044	4.8	1,434	26,760	5.4	1,095	26,284	4.2
- Other	625	18,604	3.4	386	9,583	4.0	239	9,021	2.6
- Unknown/Missing	41	1,051	3.9	20	545	3.7	21	506	4.2
NYS Excluding NYC									
- All Races	6,446	158,388	4.1	4,036	81,212	5.0	2,410	77,176	3.1
- White	5,075	127,889	4.0	3,230	65,771	4.9	1,845	62,118	3.0
- Black	1,161	24,290	4.8	682	12,222	5.6	479	12,068	4.0
- Other	182	5,500	3.3	108	2,846	3.8	74	2,654	2.8
- Unknown/Missing	28	709	3.9	16	373	4.3	12	336	3.6
New York City									
- All Races	3,896	96,759	4.0	2,304	49,546	4.7	1,592	47,213	3.4
- White	2,072	54,559	3.8	1,270	28,099	4.5	802	26,460	3.0
- Black	1,368	28,754	4.8	752	14,538	5.2	616	14,216	4.3
- Other	443	13,104	3.4	278	6,737	4.1	165	6,367	2.6
- Unknown/Missing	13	342	3.8	4	172	2.3	9	170	5.3

Section 1 - Table 1c
2000 Births - New York State Residents
Percent of Live Births with One or More Major Malformations
Sex by Race and Residence

	Both Sexes			Males			Females		
Race and Residence	Infants	Total Births	%	Infants	Total Births	%	Infants	Total Births	%
New York State									
- All Races	10,719	258,452	4.1	6,660	132,587	5.0	4,059	125,865	3.2
- White	7,333	182,284	4.0	4,605	93,455	4.9	2,728	88,829	3.1
- Black	2,660	54,497	4.9	1,584	27,862	5.7	1,076	26,635	4.0
- Other	692	20,538	3.4	450	10,704	4.2	242	9,834	2.5
- Unknown/Missing	34	1,133	3.0	21	566	3.7	13	567	2.3
NYS Excluding NYC									
- All Races	6,788	160,221	4.2	4,293	82,099	5.2	2,495	78,122	3.2
- White	5,281	127,771	4.1	3,350	65,442	5.1	1,931	62,329	3.1
- Black	1,247	25,508	4.9	763	13,067	5.8	484	12,441	3.9
- Other	234	6,073	3.9	163	3,152	5.2	71	2,921	2.4
- Unknown/Missing	26	869	3.0	17	438	3.9	9	431	2.1
New York City									
- All Races	3,931	98,231	4.0	2,367	50,488	4.7	1,564	47,743	3.3
- White	2,052	54,513	3.8	1,255	28,013	4.5	797	26,500	3.0
- Black	1,413	28,989	4.9	821	14,795	5.5	592	14,194	4.2
- Other	458	14,465	3.2	287	7,552	3.8	171	6,913	2.5
- Unknown/Missing	8	264	3.0	4	128	3.1	4	136	2.9

Section 1 - Table 1d
2001 Births - New York State Residents
Percent of Live Births with One or More Major Malformations
Sex by Race and Residence

	Both Sexes			Males			Females		
Race and Residence	Infants	Total Births	%	Infants	Total Births	%	Infants	Total Births	%
New York State									
- All Races	10,294	253,378	4.1	6,484	129,723	5.0	3,810	123,655	3.1
- White	7,129	180,517	3.9	4,529	92,335	4.9	2,600	88,182	2.9
- Black	2,485	51,812	4.8	1,539	26,442	5.8	946	25,370	3.7
- Other	639	19,998	3.2	389	10,423	3.7	250	9,575	2.6
- Unknown/Missing	41	1,051	3.9	27	523	5.2	14	528	2.7
NYS Excluding NYC									
- All Races	6,587	156,947	4.2	4,180	80,031	5.2	2,407	76,916	3.1
- White	5,137	125,230	4.1	3,279	63,803	5.1	1,858	61,427	3.0
- Black	1,176	24,806	4.7	736	12,669	5.8	440	12,137	3.6
- Other	242	6,023	4.0	146	3,134	4.7	96	2,889	3.3
- Unknown/Missing	32	888	3.6	19	425	4.5	13	463	2.8
New York City									
- All Races	3,707	96,431	3.8	2,304	49,692	4.6	1,403	46,739	3.0
- White	1,992	55,287	3.6	1,250	28,532	4.4	742	26,755	2.8
- Black	1,309	27,006	4.8	803	13,773	5.8	506	13,233	3.8
- Other	397	13,975	2.8	243	7,289	3.3	154	6,686	2.3
- Unknown/Missing	9	163	5.5	8	98	8.2	1	65	1.5

Section 1 - Table 2a
1998 Births - New York State Residents
Number of Major Malformations Per Child

Number of Malformations	Number of Children	Percent
1	8,533	78.3
2	1,495	13.7
3	482	4.4
4	178	1.6
5	97	0.9
6	46	0.4
7	29	0.3
8	19	0.2
9	11	0.1
10	5	*
11	2	*
12	2	*
>12	3	*
All Children	10,902	100.0

Section 1 - Table 2b
1999 Births - New York State Residents
Number of Major Malformations Per Child

Number of Malformations	Number of Children	Percent
1	8,208	79.4
2	1,394	13.5
3	447	4.3
4	154	1.5
5	63	0.6
6	37	0.4
7	16	0.2
8	10	0.1
9	8	0.1
10	1	*
11	3	*
>12	1	*
All Children	10,342	100.0

Section 1 - Table 2c
2000 Births - New York State Residents
Number of Major Malformations Per Child

Number of Malformations	Number of Children	Percent
1	8,786	82.0
2	1,318	12.3
3	377	3.5
4	138	1.3
5	54	0.5
6	27	0.3
7	6	0.1
8	5	*
9	1	*
10	3	*
11	1	*
12	2	*
>12	1	*
All Children	10,719	100.0

Section 1 - Table 2d
2001 Births - New York State Residents
Number of Major Malformations Per Child

Number of Malformations	Number of Children	Percent
1	8,383	81.4
2	1,306	12.7
3	367	3.6
4	139	1.4
5	58	0.6
6	25	0.2
7	8	0.1
8	7	0.1
10	1	*
All Children	10,294	100.0

* - Less than 0.05%

Note: Total percent may not add to 100% due to rounding

Section 1 - Table 3a
1998 Births - New York State Residents
Percent of Live Births with One Major Malformation
Sex by Race and Residence

	Both Sexes			Males			Females		
Race and Residence	Infants	Total Births	%	Infants	Total Births	%	Infants	Total Births	%
New York State									
- All Races	8,533	257,737	3.3	5,275	132,260	4.0	3,258	125,477	2.6
- White	5,978	184,517	3.2	3,788	94,904	4.0	2,190	89,613	2.4
- Black	2,101	54,108	3.9	1,212	27,523	4.4	889	26,585	3.3
- Other	428	17,930	2.4	260	9,208	2.8	168	8,722	1.9
- Unknown/Missing	26	1,182	2.2	15	625	2.4	11	557	2.0
NYS Excluding NYC									
- All Races	5,374	160,645	3.3	3,378	82,403	4.1	1,996	78,242	2.6
- White	4,185	129,487	3.2	2,686	66,535	4.0	1,499	62,952	2.4
- Black	1,037	25,012	4.1	607	12,766	4.8	430	12,246	3.5
- Other	135	5,357	2.5	73	2,683	2.7	62	2,674	2.3
- Unknown/Missing	17	789	2.2	12	419	2.9	5	370	1.4
New York City									
- All Races	3,159	97,092	3.3	1,897	49,857	3.8	1,262	47,235	2.7
- White	1,793	55,030	3.3	1,102	28,369	3.9	691	26,661	2.6
- Black	1,064	29,096	3.7	605	14,757	4.1	459	14,339	3.2
- Other	293	12,573	2.3	187	6,525	2.9	106	6,048	1.8
- Unknown/Missing	9	393	2.3	3	206	1.5	6	187	3.2

Section 1 - Table 3b
1999 Births - New York State Residents
Percent of Live Births with One Major Malformation
Sex by Race and Residence

	Both Sexes			Males			Females		
Race and Residence	Infants	Total Births	%	Infants	Total Births	%	Infants	Total Births	%
New York State									
- All Races	8,208	255,147	3.2	5,100	130,758	3.9	3,108	124,389	2.5
- White	5,675	182,448	3.1	3,636	93,870	3.9	2,039	88,578	2.3
- Black	1,999	53,044	3.8	1,130	26,760	4.2	869	26,284	3.3
- Other	501	18,604	2.7	318	9,583	3.3	183	9,021	2.0
- Unknown/Missing	33	1,051	3.1	16	545	2.9	17	506	3.4
NYS Excluding NYC									
- All Races	5,080	158,388	3.2	3,222	81,212	4.0	1,858	77,176	2.4
- White	4,002	127,889	3.1	2,587	65,771	3.9	1,415	62,118	2.3
- Black	917	24,290	3.8	536	12,222	4.4	381	12,068	3.2
- Other	138	5,500	2.5	86	2,846	3.0	52	2,654	2.0
- Unknown/Missing	23	709	3.2	13	373	3.5	10	336	3.0
New York City									
- All Races	3,128	96,759	3.2	1,878	49,546	3.8	1,250	47,213	2.6
- White	1,673	54,559	3.1	1,049	28,099	3.7	624	26,460	2.4
- Black	1,082	28,754	3.8	594	14,538	4.1	488	14,216	3.4
- Other	363	13,104	2.8	232	6,737	3.4	131	6,367	2.1
- Unknown/Missing	10	342	2.9	3	172	1.7	7	170	4.1

Section 1 - Table 3c
2000 Births - New York State Residents
Percent of Live Births with One Major Malformation
Sex by Race and Residence

	Both Sexes			Males			Females		
Race and Residence	Infants	Total Births	%	Infants	Total Births	%	Infants	Total Births	%
New York State									
- All Races	8,786	258,452	3.4	5,545	132,587	4.2	3,241	125,865	2.6
- White	5,997	182,284	3.3	3,823	93,455	4.1	2,174	88,829	2.4
- Black	2,194	54,497	4.0	1,334	27,862	4.8	860	26,635	3.2
- Other	568	20,538	2.8	370	10,704	3.5	198	9,834	2.0
- Unknown/Missing	27	1,133	2.4	18	566	3.2	9	567	1.6
NYS Excluding NYC									
- All Races	5,505	160,221	3.4	3,531	82,099	4.3	1,974	78,122	2.5
- White	4,297	127,771	3.4	2,763	65,442	4.2	1,534	62,329	2.5
- Black	1,000	25,508	3.9	620	13,067	4.7	380	12,441	3.1
- Other	187	6,073	3.1	134	3,152	4.3	53	2,921	1.8
- Unknown/Missing	21	869	2.4	14	438	3.2	7	431	1.6
New York City									
- All Races	3,281	98,231	3.3	2,014	50,488	4.0	1,267	47,743	2.7
- White	1,700	54,513	3.1	1,060	28,013	3.8	640	26,500	2.4
- Black	1,194	28,989	4.1	714	14,795	4.8	480	14,194	3.4
- Other	381	14,465	2.6	236	7,552	3.1	145	6,913	2.1
- Unknown/Missing	6	264	2.3	4	128	3.1	2	136	1.5

Section 1 - Table 3d
2001 Births - New York State Residents
Percent of Live Births with One Major Malformation
Sex by Race and Residence

	Both Sexes			Males			Females		
Race and Residence	Infants	Total Births	%	Infants	Total Births	%	Infants	Total Births	%
New York State									
- All Races	8,383	253,378	3.3	5,320	129,723	4.1	3,063	123,655	2.5
- White	5,785	180,517	3.2	3,708	92,335	4.0	2,077	88,182	2.4
- Black	2,057	51,812	4.0	1,275	26,442	4.8	782	25,370	3.1
- Other	502	19,998	2.5	312	10,423	3.0	190	9,575	2.0
- Unknown/Missing	39	1,051	3.7	25	523	4.8	14	528	2.7
NYS Excluding NYC									
- All Races	5,326	156,947	3.4	3,414	80,031	4.3	1,912	76,916	2.5
- White	4,152	125,230	3.3	2,679	63,803	4.2	1,473	61,427	2.4
- Black	961	24,806	3.9	604	12,669	4.8	357	12,137	2.9
- Other	182	6,023	3.0	113	3,134	3.6	69	2,889	2.4
- Unknown/Missing	31	888	3.5	18	425	4.2	13	463	2.8
New York City									
- All Races	3,057	96,431	3.2	1,906	49,692	3.8	1,151	46,739	2.5
- White	1,633	55,287	3.0	1,029	28,532	3.6	604	26,755	2.3
- Black	1,096	27,006	4.1	671	13,773	4.9	425	13,233	3.2
- Other	320	13,975	2.3	199	7,289	2.7	121	6,686	1.8
- Unknown/Missing	8	163	4.9	7	98	7.1	1	65	1.5

Section 1 - Table 4a
1998 Births - New York State Residents
Percent of Live Births with Two or More Major Malformations
Sex by Race and Residence

	Both Sexes			Males			Females		
Race and Residence	Infants	Total Births	%	Infants	Total Births	%	Infants	Total Births	%
New York State									
- All Races	2,369	257,737	0.9	1,363	132,260	1.0	1,006	125,477	0.8
- White	1,692	184,517	0.9	983	94,904	1.0	709	89,613	0.8
- Black	544	54,108	1.0	291	27,523	1.1	253	26,585	1.0
- Other	123	17,930	0.7	82	9,208	0.9	41	8,722	0.5
- Unknown/Missing	10	1,182	0.8	7	625	1.1	3	557	0.5
NYS Excluding NYC									
- All Races	1,507	160,645	0.9	885	82,403	1.1	622	78,242	0.8
- White	1,206	129,487	0.9	710	66,535	1.1	496	62,952	0.8
- Black	247	25,012	1.0	137	12,766	1.1	110	12,246	0.9
- Other	48	5,357	0.9	33	2,683	1.2	15	2,674	0.6
- Unknown/Missing	6	789	0.8	5	419	1.2	1	370	0.3
New York City									
- All Races	862	97,092	0.9	478	49,857	1.0	384	47,235	0.8
- White	486	55,030	0.9	273	28,369	1.0	213	26,661	0.8
- Black	297	29,096	1.0	154	14,757	1.0	143	14,339	1.0
- Other	75	12,573	0.6	49	6,525	0.8	26	6,048	0.4
- Unknown/Missing	4	393	1.0	2	206	1.0	2	187	1.1

Section 1 - Table 4b
1999 Births - New York State Residents
Percent of Live Births with Two or More Major Malformations
Sex by Race and Residence

	Both Sexes			Males			Females		
Race and Residence	Infants	Total Births	%	Infants	Total Births	%	Infants	Total Births	%
New York State									
- All Races	2,134	255,147	0.8	1,240	130,758	0.9	894	124,389	0.7
- White	1,472	182,448	0.8	864	93,870	0.9	608	88,578	0.7
- Black	530	53,044	1.0	304	26,760	1.1	226	26,284	0.9
- Other	124	18,604	0.7	68	9,583	0.7	56	9,021	0.6
- Unknown/Missing	8	1,051	0.8	4	545	0.7	4	506	0.8
NYS Excluding NYC									
- All Races	1,366	158,388	0.9	814	81,212	1.0	552	77,176	0.7
- White	1,073	127,889	0.8	643	65,771	1.0	430	62,118	0.7
- Black	244	24,290	1.0	146	12,222	1.2	98	12,068	0.8
- Other	44	5,500	0.8	22	2,846	0.8	22	2,654	0.8
- Unknown/Missing	5	709	0.7	3	373	0.8	2	336	0.6
New York City									
- All Races	768	96,759	0.8	426	49,546	0.9	342	47,213	0.7
- White	399	54,559	0.7	221	28,099	0.8	178	26,460	0.7
- Black	286	28,754	1.0	158	14,538	1.1	128	14,216	0.9
- Other	80	13,104	0.6	46	6,737	0.7	34	6,367	0.5
- Unknown/Missing	3	342	0.9	1	172	0.6	2	170	1.2

Section 1 - Table 4c
2000 Births - New York State Residents
Percent of Live Births with Two or More Major Malformations
Sex by Race and Residence

	Both Sexes			Males			Females		
Race and Residence	Infants	Total Births	%	Infants	Total Births	%	Infants	Total Births	%
New York State									
- All Races	1,934	258,452	0.7	1,115	132,587	0.8	819	125,865	0.7
- White	1,337	182,284	0.7	782	93,455	0.8	555	88,829	0.6
- Black	466	54,497	0.9	250	27,862	0.9	216	26,635	0.8
- Other	124	20,538	0.6	80	10,704	0.7	44	9,834	0.4
- Unknown/Missing	7	1,133	0.6	3	566	0.5	4	567	0.7
NYS Excluding NYC									
- All Races	1,283	160,221	0.8	762	82,099	0.9	521	78,122	0.7
- White	984	127,771	0.8	587	65,442	0.9	397	62,329	0.6
- Black	247	25,508	1.0	143	13,067	1.1	104	12,441	0.8
- Other	47	6,073	0.8	29	3,152	0.9	18	2,921	0.6
- Unknown/Missing	5	869	0.6	3	438	0.7	2	431	0.5
New York City									
- All Races	651	98,231	0.7	353	50,488	0.7	298	47,743	0.6
- White	353	54,513	0.6	195	28,013	0.7	158	26,500	0.6
- Black	219	28,989	0.8	107	14,795	0.7	112	14,194	0.8
- Other	77	14,465	0.5	51	7,552	0.7	26	6,913	0.4
- Unknown/Missing	2	264	0.8	0	128	0.0	2	136	1.5

Section 1 - Table 4d
2001 Births - New York State Residents
Percent of Live Births with Two or More Major Malformations
Sex by Race and Residence

	Both Sexes			Males			Females		
Race and Residence	Infants	Total Births	%	Infants	Total Births	%	Infants	Total Births	%
New York State									
- All Races	1,911	253,378	0.8	1,164	129,723	0.9	747	123,655	0.6
- White	1,344	180,517	0.7	821	92,335	0.9	523	88,182	0.6
- Black	428	51,812	0.8	264	26,442	1.0	164	25,370	0.6
- Other	137	19,998	0.7	77	10,423	0.7	60	9,575	0.6
- Unknown/Missing	2	1,051	0.2	2	523	0.4	0	528	0.0
NYS Excluding NYC									
- All Races	1,261	156,947	0.8	766	80,031	1.0	495	76,916	0.6
- White	985	125,230	0.8	600	63,803	0.9	385	61,427	0.6
- Black	215	24,806	0.9	132	12,669	1.0	83	12,137	0.7
- Other	60	6,023	1.0	33	3,134	1.1	27	2,889	0.9
- Unknown/Missing	1	888	0.1	1	425	0.2	0	463	0.0
New York City									
- All Races	650	96,431	0.7	398	49,692	0.8	252	46,739	0.5
- White	359	55,287	0.6	221	28,532	0.8	138	26,755	0.5
- Black	213	27,006	0.8	132	13,773	1.0	81	13,233	0.6
- Other	77	13,975	0.6	44	7,289	0.6	33	6,686	0.5
- Unknown/Missing	1	163	0.6	1	98	1.0	0	65	0.0

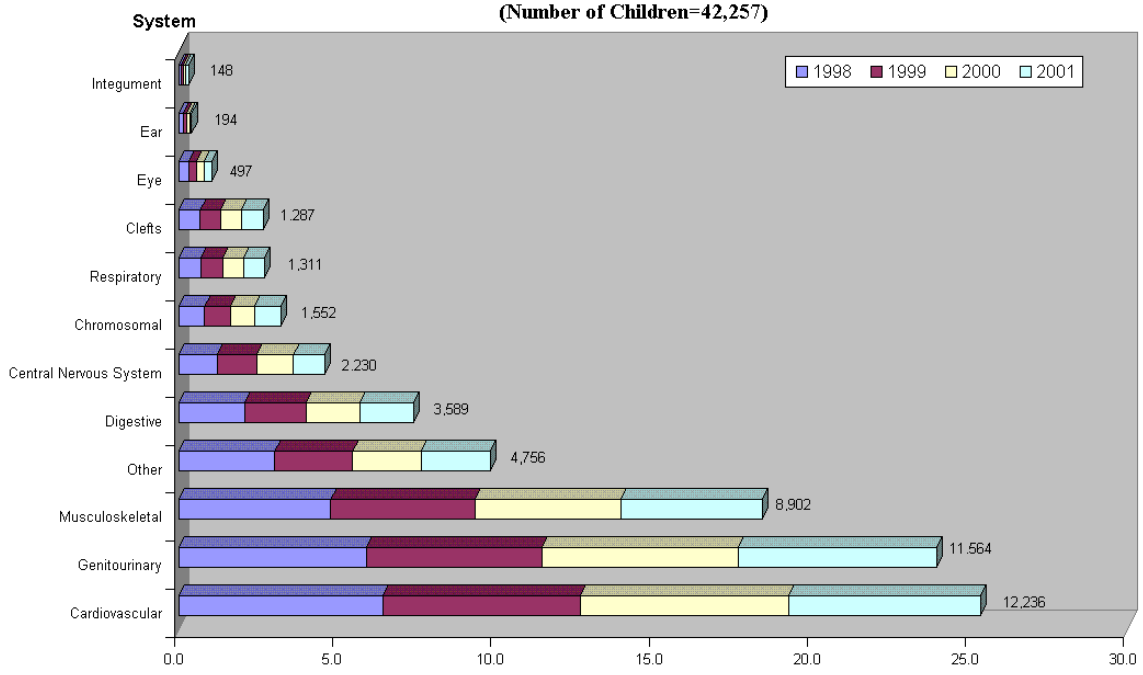
Section II
Major Congenital Malformations by Organ System, 1998-2001

Introduction to Figures

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

Major Malformations by Organ System
1998 to 2001 Births - New York State Residents

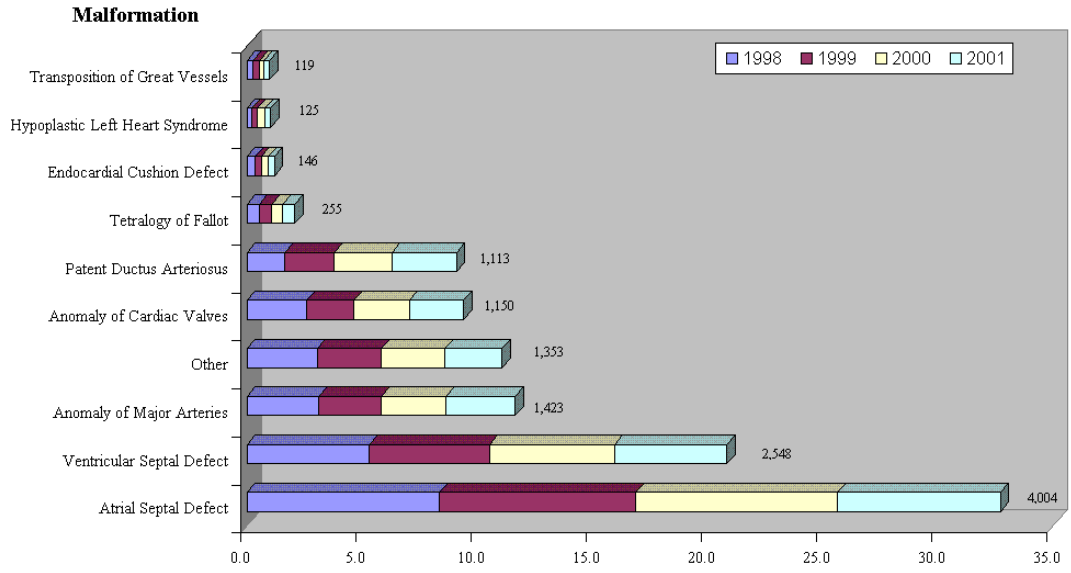
(Number of Children=42,257)



Percent of total number of malformations

Major Malformations by Organ System
1998 to 2001 Births - New York State Residents

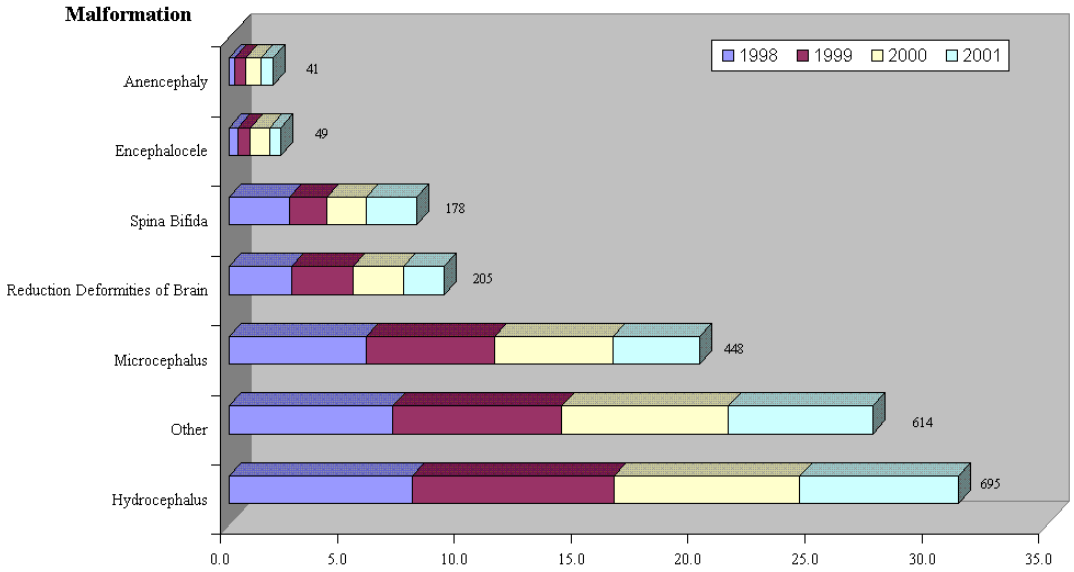
Cardiovascular System Subset Category
(Number of Children=10,512)



Percent of total number of malformations for subset

Major Malformations by Organ System
1998 to 2001 Births - New York State Residents

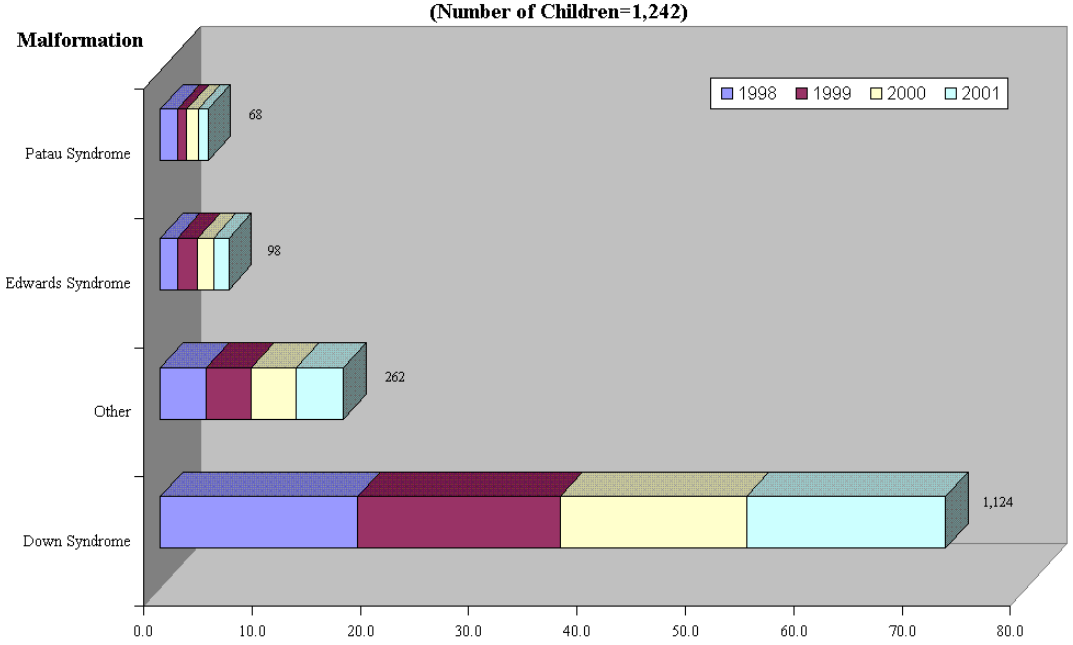
Central Nervous System Subset Category
(Number of Children=1,611)



Percent of total number of malformations for subset

Major Malformations by Organ System
1998 to 2001 Births - New York State Residents

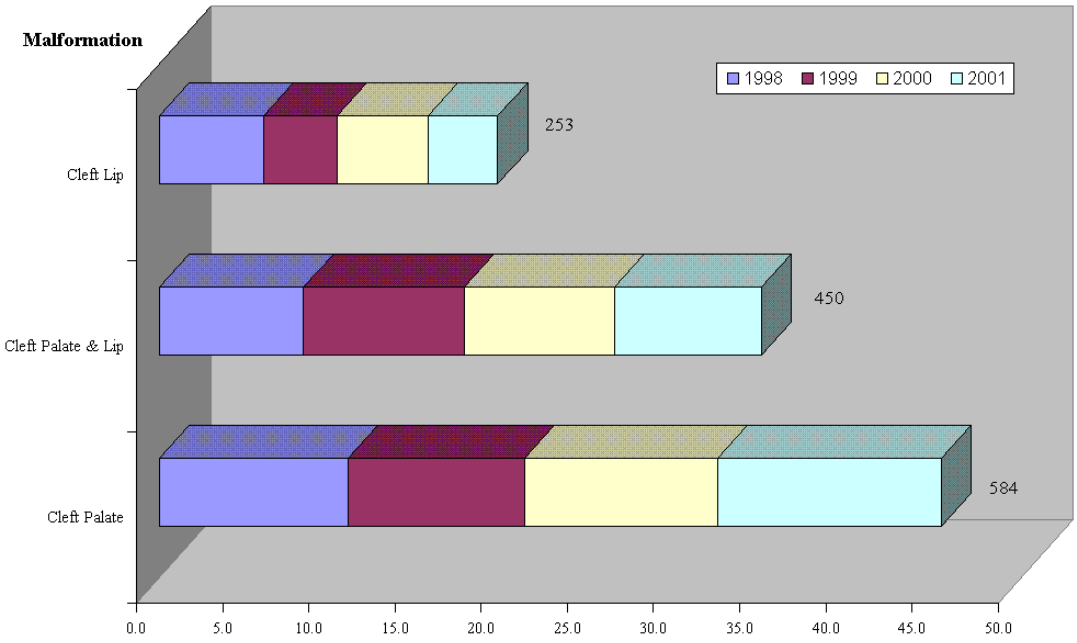
Chromosomal Subset Category
(Number of Children=1,242)



Percent of total number of malformations for subset

Major Malformations by Organ System
1998 to 2001 Births - New York State Residents

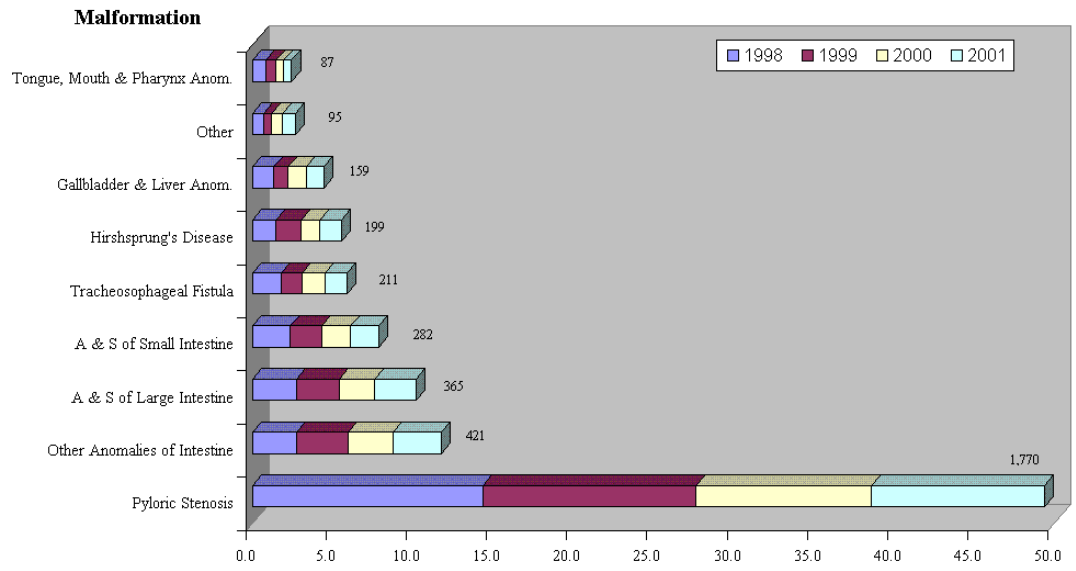
Oral Clefts Subset Category
(Number of Children=1,061)



Percent of total number of malformations for subset

Major Malformations by Organ System
1998 to 2001 Births - New York State Residents

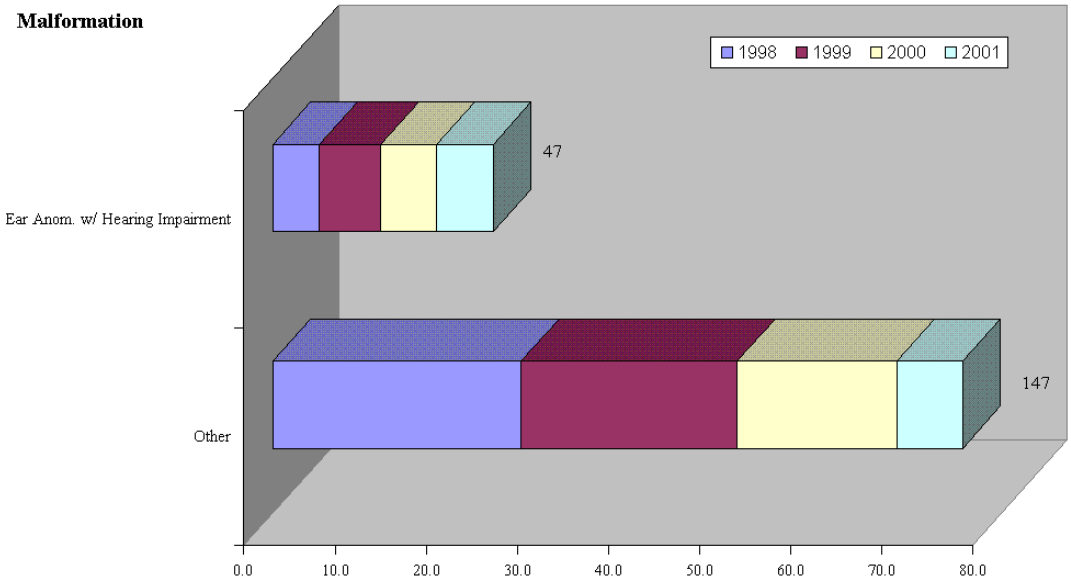
Digestive System Subset Category
(Number of Children=3,109)



Percent of total number of malformations for subset

Major Malformations by Organ System
1998 to 2001 Births - New York State Residents

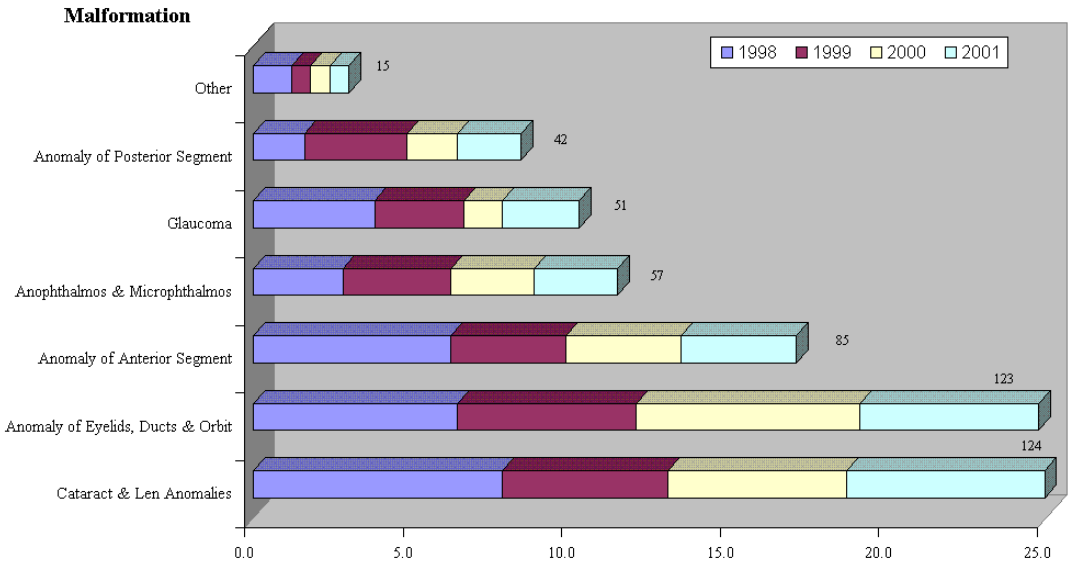
Ear Subset Category
(Number of Children=126)



Percent of total number of malformations for subset

Major Malformations by Organ System
1998 to 2001 Births - New York State Residents

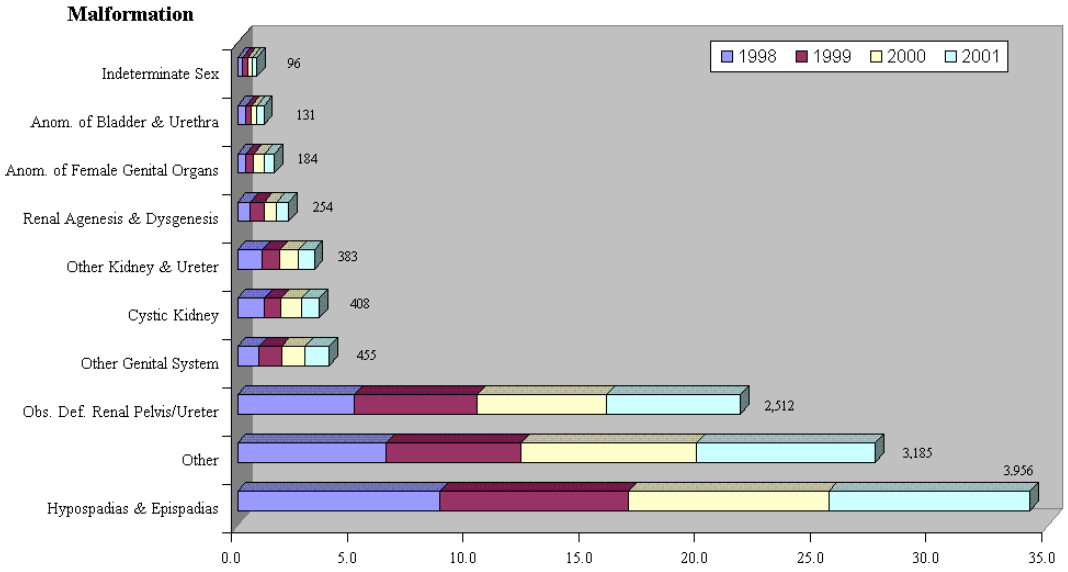
Eye Subset Category
(Number of Children=367)



Percent of total number of malformations for subset

Major Malformations by Organ System
1998 to 2001 Births - New York State Residents

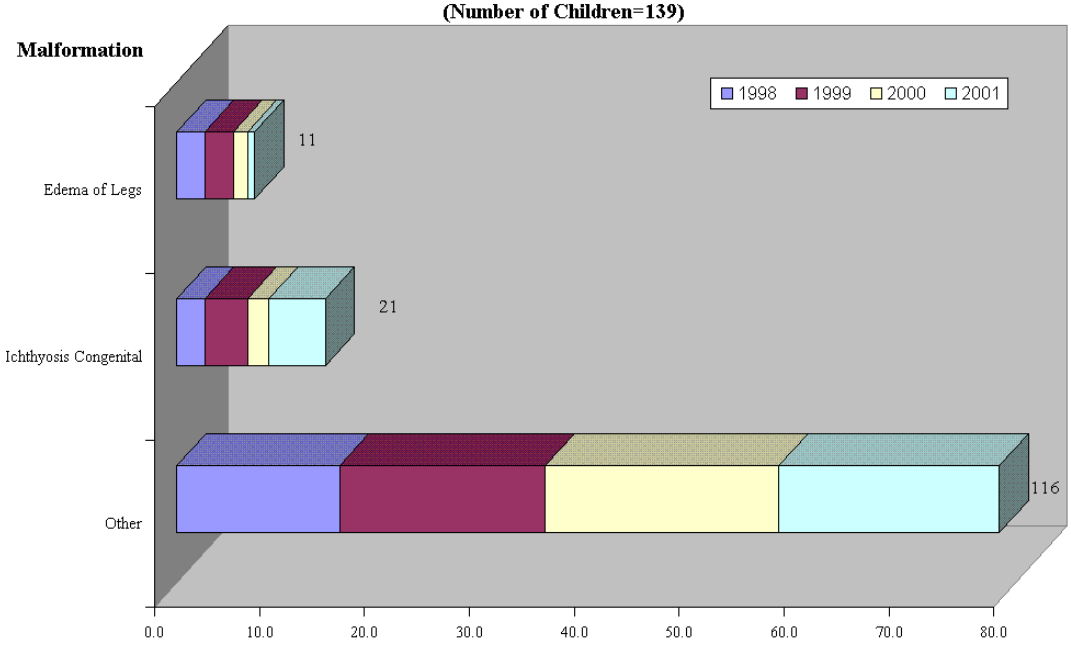
Genitourinary System Subset Category
(Number of Children=10,811)



Percent of total number of malformations for subset

Major Malformations by Organ System
1998 to 2001 Births - New York State Residents

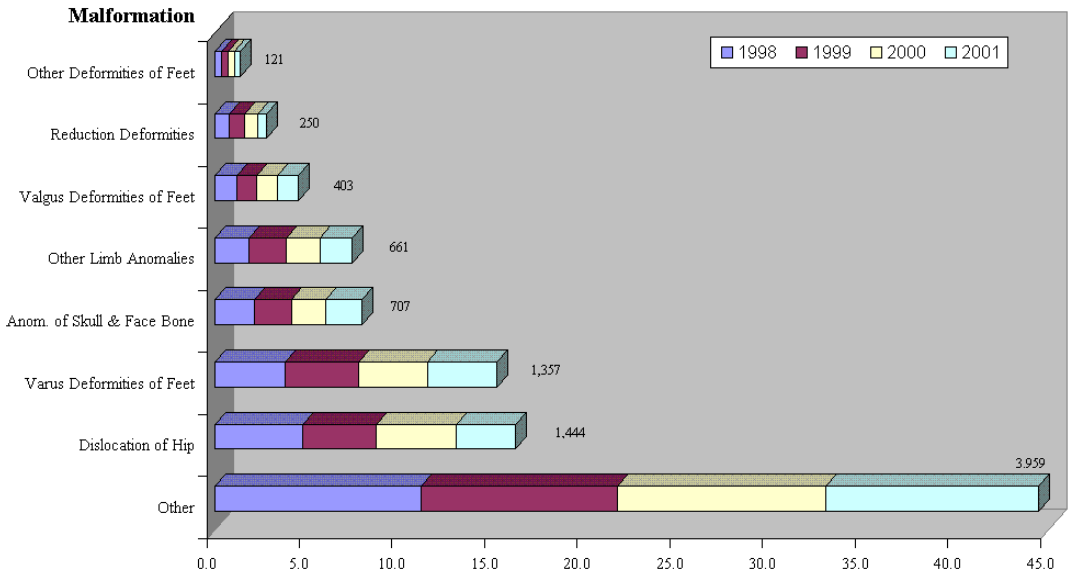
Integument System Subset Category
(Number of Children=139)



Percent of total number of malformations for subset

Major Malformations by Organ System
1998 to 2001 Births - New York State Residents

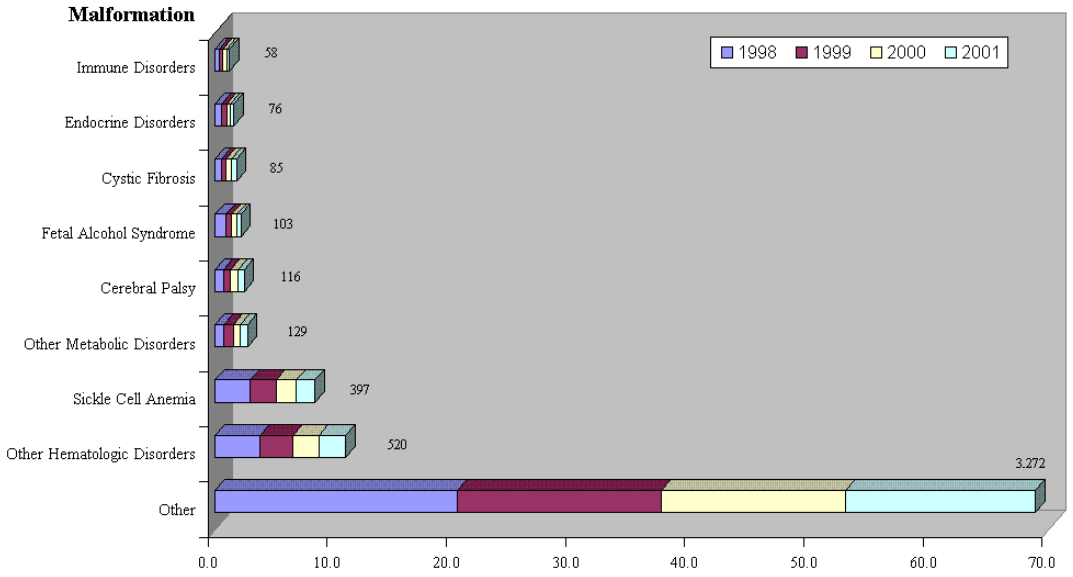
Musculoskeletal System Subset Category
(Number of Children=8,311)



Percent of total number of malformations for subset

Major Malformations by Organ System
1998 to 2001 Births - New York State Residents

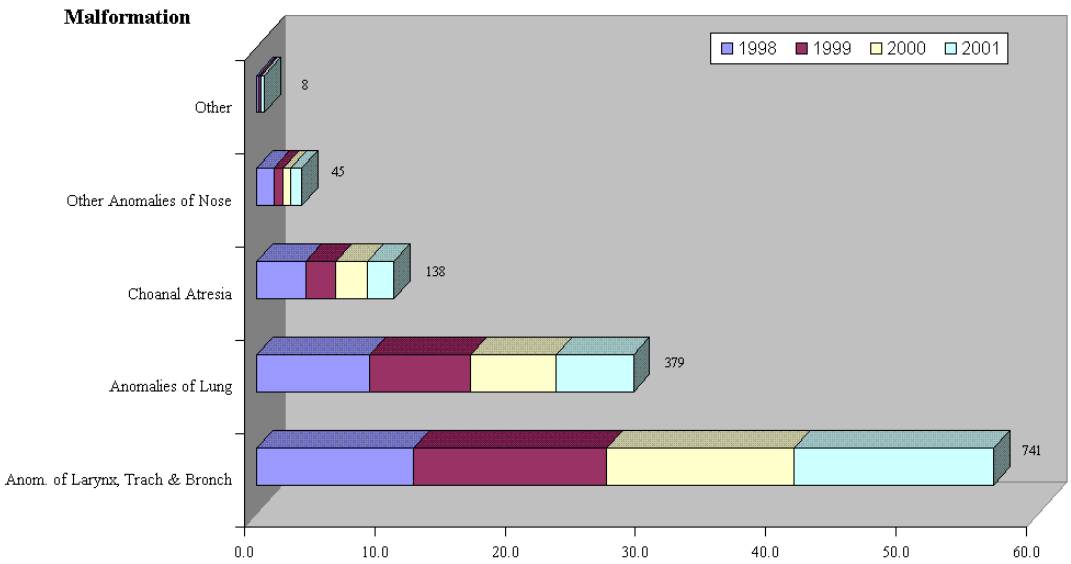
Other Subset Category
(Number of Children=3,994)



Percent of total number of malformations for subset

Major Malformations by Organ System
1998 to 2001 Births - New York State Residents

Respiratory System Subset Category
(Number of Children=974)



Percent of total number of malformations for subset

Section III
Prevalence of Selected Malformations By Sex and Race

Introduction to Tables

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

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

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

1998 Births– New York State Residents

ICD-9 Code	Malformation	Total Number	Total Prevalence	Male	Female	Ratio (M/F)	White	Black	Other
243	Congenital hypothyroidism	54	2.1	2.2	2.0	1.1	2.0	2.2	2.6
270.1	Phenylketonuria	2	0.1	0.0	0.2	0.0	0.1	0.2	0.0
277.0	Cystic fibrosis	27	1.0	1.0	1.1	0.9	1.2	0.7	0.0
282.6	Sickle-cell anemia	142	5.5	5.8	5.2	1.1	0.7	23.8	0.5
740.0	Anencephalus	5	0.2	0.2	0.2	0.6	0.1	0.6	0.0
741.0	Spina bifida with hydrocephalus	25	1.0	0.9	1.0	0.9	0.9	1.5	0.0
741.9	Spina bifida without hydrocephalus	43	1.7	1.8	1.5	1.2	1.7	1.8	0.5
742.0	Encephalocele	10	0.4	0.2	0.6	0.2	0.4	0.4	0.0
742.1	Microcephalus	141	5.5	4.2	6.8	0.6	4.2	10.0	4.7
742.2	Agyria & lissencephaly	8	0.3	0.5	0.2	2.8	0.3	0.2	0.5
742.2	Anomalies of corpus callosum	37	1.4	1.7	1.2	1.4	1.6	0.6	2.1
742.2	Holoprosencephaly	15	0.6	0.5	0.7	0.6	0.5	0.9	0.0
742.3	Congenital hydrocephalus	193	7.5	8.2	6.8	1.2	6.4	10.7	8.4
742.4	Porencephaly	7	0.3	0.3	0.2	1.3	0.2	0.6	0.5
742.5	Congenital tethered cord	20	0.8	0.8	0.8	0.9	0.9	0.4	0.5
743.0	Anophthalmos	3	0.1	0.1	0.2	0.5	0.1	0.2	0.0
743.1	Microphthalmos	17	0.7	0.7	0.6	1.1	0.8	0.4	0.5
743.2	Glaucoma	21	0.8	0.8	0.8	1.0	0.5	1.3	2.1
743.3	Absence of lens	3	0.1	0.2	0.1	1.9	0.1	0.2	0.0
743.3	Congenital cataract	36	1.4	1.4	1.4	0.9	1.5	1.7	0.0
743.45	Aniridia	4	0.2	0.1	0.2	0.3	0.2	0.0	0.0
743.46	Coloboma of iris	4	0.2	0.2	0.1	2.8	0.2	0.0	0.5
744.0	Anotia/microtia	20	0.8	1.0	0.6	1.8	0.9	0.2	1.0
745.0	Common truncus	14	0.5	0.5	0.6	0.7	0.5	0.7	0.0
745.1	Transposition of great vessels	100	3.9	5.0	2.7	1.8	4.0	3.5	3.7
745.2	Tetralogy of Fallot	127	4.9	5.3	4.5	1.2	4.5	5.4	6.3
745.3	Common ventricle	20	0.8	0.8	0.8	0.9	0.8	0.6	1.6
745.4	Ventricular septal defect	935	36.3	34.2	38.5	0.9	38.7	31.8	25.6
745.5	Ostium secundum type atrial septal def.	1,340	52.0	51.1	52.9	1.0	48.0	70.4	36.6
745.6	Endocardial cushion defects	75	2.9	2.6	3.2	0.8	3.3	2.4	1.0
746.0	Atresia/stenosis of pulmonary valve	244	9.5	9.8	9.2	1.1	8.5	13.7	5.8
746.1	Tricuspid atresia/stenosis/hypoplasia	24	0.9	1.1	0.8	1.3	0.8	1.3	1.0
746.2	Ebstein's anomaly	17	0.7	1.0	0.3	3.1	0.8	0.2	0.5
746.3	Congenital stenosis of aortic valve	31	1.2	2.0	0.4	4.9	1.4	0.9	0.5
746.7	Hypoplastic left heart syndrome	42	1.6	1.7	1.6	1.0	1.8	0.9	1.6
746.85	Anomalies of coronary artery	15	0.6	0.8	0.3	2.6	0.5	0.9	0.5
747.0	Patent ductus arteriosis	207	8.0	8.2	7.8	1.1	6.6	12.9	6.8
747.10	Coartation of aorta	92	3.6	4.3	2.8	1.5	3.9	2.6	3.1
747.41	Total anomalous pulmonary venus connect.	25	1.0	1.4	0.5	3.0	1.1	0.6	1.0

1998 Births - New York State Residents (continued)

ICD-9 Code	Malformation	Total Number	Total Prevalence	Male	Female	Ratio (M/F)	White	Black	Other
748.0	Choanal atresia	51	2.0	1.9	2.1	0.9	2.4	0.6	1.6
748.5	Agenesis/hypoplasia of lung	94	3.6	3.9	3.3	1.2	3.7	3.7	2.6
749.0	Cleft palate	144	5.6	4.6	6.6	0.7	6.4	3.7	2.6
749.1	Cleft lip	83	3.2	3.6	2.9	1.2	3.7	1.5	2.6
749.2	Cleft palate & lip	111	4.3	4.5	4.1	1.1	4.6	2.8	5.8
750.3	Tracheoesophageal fistula etc.	71	2.8	2.9	2.6	1.2	3.5	1.1	0.0
750.5	Congenital hypertrophic pyloric stenosis	518	20.1	31.8	7.7	4.1	24.3	10.5	6.3
751.1	Atresia and stenosis of small intestine	98	3.8	3.2	4.5	0.7	3.9	3.7	2.6
751.2	Atresia and stenosis of rectum or anus	109	4.2	4.8	3.6	1.3	4.4	4.3	2.1
751.3	Hirschsprungs disease	57	2.2	2.6	1.8	1.5	2.3	1.3	3.7
751.4	Anomalies of intestinal fixation	47	1.8	2.2	1.4	1.5	1.6	3.1	0.0
751.61	Biliary atresia	31	1.2	0.8	1.6	0.5	0.8	2.2	2.1
752.6	Epispadias	17	0.7	1.3	0.0		0.7	0.9	0.0
752.6	Hypospadias	894	34.7	67.3	0.3	211.1	38.3	26.8	20.9
753.0	Renal agenesis and dysgenesis	83	3.2	3.6	2.9	1.2	3.0	4.1	2.6
753.1	Cystic kidney disease	163	6.3	7.7	4.9	1.6	5.7	7.8	7.8
753.2	Obstructive defect renal pelvis & ureter	621	24.1	32.9	14.8	2.2	25.7	18.5	23.5
753.5	Extrophy of urinary bladder	8	0.3	0.4	0.2	1.6	0.3	0.2	0.0
753.6	Atresia & stenosis of urethra & bladder	21	0.8	1.5	0.1	19.0	0.8	0.6	1.6
754.3	Congenital dislocation of hip	332	12.9	5.2	21.0	0.2	15.0	5.5	13.6
754.51	Talipes equinovarus	257	10.0	12.5	7.3	1.7	10.7	9.4	3.1
755.2	Reduction deformities of upper limb	57	2.2	1.7	2.7	0.6	2.5	1.7	0.5
755.3	Reduction deformities of lower limb	42	1.6	1.4	1.8	0.8	1.5	1.7	2.1
755.8	Arthrogryposis multiplex congenita	26	1.0	1.0	1.0	0.9	1.0	1.5	0.0
756.0	Craniosynostosis	90	3.5	4.8	2.2	2.2	4.1	2.2	1.0
756.0	Goldenhar syndrome	16	0.6	0.8	0.5	1.6	0.6	0.7	0.5
756.4	Chonodrodystrophy	21	0.8	1.1	0.5	2.4	0.7	1.5	0.0
756.51	Osteogenesis imperfecta	13	0.5	0.4	0.6	0.6	0.5	0.6	0.0
756.6	Diaphragmatic hernia	53	2.1	2.6	1.5	1.7	2.2	1.8	1.6
756.7	Gastroschisis	18	0.7	0.9	0.5	1.9	0.7	0.9	0.0
756.7	Omphalocele	25	1.0	0.9	1.0	0.9	1.1	0.4	1.0
756.7	Prune belly	5	0.2	0.4	0.0		0.2	0.4	0.0
758.0	Down syndrome	282	10.9	11.6	10.2	1.1	12.0	7.4	10.5
758.1	Patau syndrome	24	0.9	0.8	1.1	0.7	0.9	0.9	1.0
758.2	Edwards syndrome	24	0.9	0.8	1.1	0.7	0.6	1.8	1.6
758.6	Gonadal dysgenesis	7	0.3	0.1	0.5	0.2	0.4	0.0	0.0
758.7	Klinefelter syndrome	13	0.5	0.9	0.1	11.4	0.7	0.0	0.5
759.3	Situs inversus	21	0.8	0.9	0.7	1.3	0.7	0.9	1.6
760.71	Fetal alcohol syndrome	45	1.7	1.7	1.8	0.9	0.7	6.1	0.0
771.0	Congenital rubella	2	0.1	0.2	0.0		0.1	0.2	0.0
771.1	Congenital cytomegalovirus infection	25	1.0	1.0	1.0	1.0	0.5	2.4	1.0
771.2	Other congenital infections	35	1.4	1.5	1.2	1.3	1.2	2.0	0.5

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

1999 Births– New York State Residents

ICD-9 Code	Malformation	Total Number	Total Prevalence	Male	Female	Ratio (M/F)	White	Black	Other
243	Congenital hypothyroidism	38	1.5	1.9	1.0	1.8	1.3	2.1	1.5
270.1	Phenylketonuria	3	0.1	0.1	0.2	0.5	0.1	0.2	0.0
277.0	Cystic fibrosis	19	0.7	0.8	0.6	1.3	0.7	0.9	0.0
282.6	Sickle-cell anemia	103	4.0	4.7	3.4	1.4	0.3	18.1	0.0
740.0	Anencephalus	11	0.4	0.4	0.5	0.8	0.4	0.4	0.0
741.0	Spina bifida with hydrocephalus	19	0.7	0.5	1.0	0.4	0.7	0.8	1.0
741.9	Spina bifida without hydrocephalus	26	1.0	1.1	1.0	1.1	1.1	0.8	1.0
742.0	Encephalocele	15	0.6	0.6	0.6	1.1	0.6	0.6	0.5
742.1	Microcephalus	127	5.0	4.1	5.9	0.7	4.3	7.9	3.6
742.2	Agyria & lissencephaly	4	0.2	0.2	0.2	1.0	0.2	0.0	0.0
742.2	Anomalies of corpus callosum	45	1.8	1.8	1.7	1.1	1.9	1.9	0.5
742.2	Holoprosencephaly	10	0.4	0.5	0.2	2.2	0.4	0.6	0.0
742.3	Congenital hydrocephalus	209	8.2	9.6	6.8	1.4	6.9	13.6	5.6
742.4	Porencephaly	5	0.2	0.2	0.2	1.4	0.2	0.0	0.5
742.5	Congenital tethered cord	22	0.9	0.9	0.8	1.1	1.2	0.2	0.0
743.1	Microphthalmos	22	0.9	0.8	1.0	0.8	0.9	0.9	0.5
743.2	Glaucoma	15	0.6	0.8	0.4	1.9	0.5	0.8	0.5
743.3	Absence of lens	2	0.1	0.2	0.0		0.1	0.0	0.0
743.3	Congenital cataract	24	0.9	1.1	0.7	1.6	1.0	0.9	0.5
743.45	Aniridia	1	0.0	0.1	0.0		0.0	0.0	0.5
743.46	Coloboma of iris	2	0.1	0.1	0.1	1.0	0.1	0.0	0.0
744.0	Anotia/microtia	20	0.8	0.8	0.7	1.2	0.9	0.4	0.5
745.0	Common truncus	19	0.7	0.8	0.7	1.1	0.8	0.9	0.0
745.1	Transposition of great vessels	103	4.0	5.0	3.0	1.7	4.3	2.8	4.6
745.2	Tetralogy of Fallot	106	4.2	4.6	3.7	1.2	3.9	5.5	3.1
745.3	Common ventricle	19	0.7	0.7	0.8	0.9	0.9	0.4	0.5
745.4	Ventricular septal defect	870	34.1	31.7	36.6	0.9	34.9	30.9	34.1
745.5	Ostium secundum type atrial septal def.	1,308	51.3	51.9	50.6	1.0	45.1	78.4	32.0
745.6	Endocardial cushion defects	67	2.6	2.2	3.1	0.7	2.6	3.0	1.5
746.0	Atresia/stenosis of pulmonary valve	198	7.8	7.6	7.9	1.0	6.4	14.3	3.1
746.1	Tricuspid atresia/stenosis/hypoplasia	23	0.9	0.7	1.1	0.6	0.9	0.9	1.0
746.2	Ebstein's anomaly	8	0.3	0.3	0.3	1.0	0.4	0.0	0.5
746.3	Congenital stenosis of aortic valve	17	0.7	1.0	0.3	3.1	0.8	0.0	1.0
746.7	Hypoplastic left heart syndrome	42	1.6	2.1	1.2	1.7	1.8	1.9	0.0
746.85	Anomalies of coronary artery	8	0.3	0.5	0.2	2.9	0.1	1.1	0.0
747.0	Patent ductus arteriosis	270	10.6	9.2	12.1	0.8	9.8	14.5	7.1
747.10	Coartation of aorta	119	4.7	5.0	4.3	1.2	4.4	4.9	5.6
747.41	Total anomalous pulmonary venus connect.	22	0.9	1.1	0.6	1.7	1.0	0.4	0.0
748.0	Choanal atresia	30	1.2	1.3	1.0	1.2	1.3	0.9	1.0

1999 Births – New York State Residents (continued)

ICD-9 Code	Malformation	Total Number	Total Prevalence	Male	Female	Ratio (M/F)	White	Black	Other
748.5	Agenesis/hypoplasia of lung	80	3.1	2.9	3.4	0.9	2.6	4.9	2.0
749.0	Cleft palate	135	5.3	5.4	5.2	1.0	5.3	4.3	7.1
749.1	Cleft lip	56	2.2	3.0	1.4	2.2	2.4	1.1	3.1
749.2	Cleft palate & lip	120	4.7	6.0	3.3	1.8	5.1	3.4	4.6
750.3	Tracheoesophageal fistula etc.	51	2.0	2.1	1.9	1.1	2.3	1.3	1.0
750.5	Congenital hypertrophic pyloric stenosis	478	18.7	29.4	7.5	3.9	22.4	10.9	6.1
751.1	Atresia and stenosis of small intestine	78	3.1	3.2	2.9	1.1	2.6	4.1	4.1
751.2	Atresia and stenosis of rectum or anus	104	4.1	4.7	3.5	1.3	4.3	2.8	5.1
751.3	Hirschsprungs disease	59	2.3	3.2	1.4	2.4	1.9	3.2	3.6
751.4	Anomalies of intestinal fixation	54	2.1	2.6	1.6	1.6	2.1	2.6	1.0
751.61	Biliary atresia	23	0.9	1.3	0.5	2.7	0.7	1.5	1.5
752.6	Epispadias	8	0.3	0.5	0.1	6.7	0.3	0.4	0.0
752.6	Hypospadias	822	32.2	62.5	0.4	155.4	34.5	27.1	22.4
753.0	Renal agenesis and dysgenesis	76	3.0	3.7	2.2	1.7	2.7	3.4	3.1
753.1	Cystic kidney disease	118	4.6	5.4	3.9	1.4	4.2	6.8	2.0
753.2	Obstructive defect renal pelvis & ureter	654	25.6	34.7	16.1	2.2	27.1	20.7	23.4
753.5	Extrophy of urinary bladder	6	0.2	0.2	0.3	0.5	0.3	0.0	0.0
753.6	Atresia & stenosis of urethra & bladder	19	0.7	1.4	0.1	17.1	0.5	1.3	0.5
754.3	Congenital dislocation of hip	286	11.2	7.1	15.5	0.5	12.2	5.7	16.8
754.51	Talipes equinovarus	269	10.5	12.8	8.1	1.6	11.3	9.8	5.1
755.2	Reduction deformities of upper limb	48	1.9	2.3	1.4	1.6	2.2	1.1	1.0
755.3	Reduction deformities of lower limb	32	1.3	1.1	1.4	0.7	1.3	1.5	0.0
755.8	Arthrogryposis multiplex congenita	10	0.4	0.5	0.2	2.2	0.5	0.2	0.0
756.0	Craniosynostosis	96	3.8	4.8	2.7	1.8	4.2	3.0	2.0
756.0	Goldenhar syndrome	12	0.5	0.5	0.5	1.0	0.4	0.6	0.5
756.4	Chonodrodystrophy	30	1.2	0.8	1.6	0.5	1.1	1.9	0.0
756.51	Osteogenesis imperfecta	16	0.6	0.5	0.7	0.7	0.8	0.4	0.0
756.6	Diaphragmatic hernia	52	2.0	2.3	1.8	1.3	2.0	2.3	1.5
756.7	Gastroschisis	43	1.7	1.1	2.3	0.5	1.7	2.3	0.0
756.7	Omphalocele	19	0.7	0.3	1.2	0.3	0.8	0.9	0.0
756.7	Prune belly	5	0.2	0.2	0.2	1.4	0.2	0.2	0.0
758.0	Down syndrome	291	11.4	11.3	11.5	1.0	12.6	10.0	4.6
758.1	Patau syndrome	14	0.5	0.7	0.4	1.7	0.5	0.8	0.0
758.2	Edwards syndrome	30	1.2	0.8	1.5	0.6	1.0	1.7	1.0
758.6	Gonadal dysgenesis	12	0.5	0.1	0.9	0.1	0.4	0.8	0.0
758.7	Klinefelter syndrome	17	0.7	1.3	0.0		0.7	0.8	0.5
759.3	Situs inversus	18	0.7	0.9	0.5	1.9	0.7	0.4	2.0
760.71	Fetal alcohol syndrome	25	1.0	1.4	0.6	2.4	0.4	2.8	0.5
771.0	Congenital rubella	3	0.1	0.1	0.2	0.5	0.1	0.4	0.0
771.1	Congenital cytomegalovirus infection	15	0.6	0.7	0.5	1.4	0.5	1.1	0.0
771.2	Other congenital infections	39	1.5	1.7	1.4	1.2	1.4	2.3	0.5

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

2000 Births– New York State Residents

ICD-9 Code	Malformation	Total Number	Total Prevalence	Male	Female	Ratio (M/F)	White	Black	Other
243	Congenital hypothyroidism	52	2.0	2.4	1.6	1.5	1.5	3.3	2.8
270.1	Phenylketonuria	1	0.0	0.0	0.1	0.0	0.1	0.0	0.0
277.0	Cystic fibrosis	20	0.8	0.7	0.9	0.8	1.0	0.4	0.0
282.6	Sickle-cell anemia	82	3.2	3.2	3.1	1.0	0.4	13.6	0.0
740.0	Anencephalus	15	0.6	0.8	0.4	1.9	0.7	0.2	0.9
741.0	Spina bifida with hydrocephalus	20	0.8	0.8	0.8	0.9	0.7	1.1	0.0
741.9	Spina bifida without hydrocephalus	23	0.9	1.0	0.8	1.2	0.8	1.3	0.9
742.0	Encephalocele	25	1.0	0.9	1.0	0.9	0.8	1.7	0.9
742.1	Microcephalus	118	4.6	4.1	5.0	0.8	3.3	7.7	6.9
742.2	Agyria & lissencephaly	2	0.1	0.2	0.0		0.1	0.2	0.0
742.2	Anomalies of corpus callosum	29	1.1	1.3	1.0	1.3	1.3	1.1	0.0
742.2	Holoprosencephaly	16	0.6	0.3	1.0	0.3	0.7	0.7	0.0
742.3	Congenital hydrocephalus	191	7.4	8.1	6.6	1.2	6.3	11.7	6.0
742.4	Porencephaly	10	0.4	0.5	0.2	2.2	0.5	0.2	0.0
742.5	Congenital tethered cord	28	1.1	1.2	1.0	1.3	1.3	0.6	0.5
743.1	Microphthalmos	15	0.6	0.7	0.5	1.4	0.6	0.6	0.5
743.2	Glaucoma	6	0.2	0.2	0.3	0.5	0.2	0.4	0.0
743.3	Absence of lens	1	0.0	0.0	0.1	0.0	0.1	0.0	0.0
743.3	Congenital cataract	29	1.1	0.8	1.4	0.6	1.1	1.3	0.9
743.46	Coloboma of iris	4	0.2	0.1	0.2	0.3	0.1	0.4	0.0
744.0	Anotia/microtia	13	0.5	0.7	0.3	2.1	0.5	0.2	0.9
745.0	Common truncus	13	0.5	0.3	0.7	0.4	0.5	0.4	0.5
745.1	Transposition of great vessels	82	3.2	3.6	2.7	1.3	3.2	2.6	4.6
745.2	Tetralogy of Fallot	89	3.4	4.4	2.4	1.9	3.2	3.1	6.5
745.3	Common ventricle	7	0.3	0.5	0.1	5.7	0.2	0.6	0.0
745.4	Ventricular septal defect	894	34.6	31.3	38.0	0.8	35.4	31.9	32.3
745.5	Ostium secundum type atrial septal def.	1,366	52.8	54.8	50.7	1.1	48.5	71.9	39.2
745.6	Endocardial cushion defects	52	2.0	2.2	1.8	1.2	1.9	2.8	1.4
746.0	Atresia/stenosis of pulmonary valve	206	8.0	7.2	8.8	0.8	7.5	10.3	6.0
746.1	Tricuspid atresia/stenosis/hypoplasia	17	0.7	0.8	0.5	1.7	0.4	1.3	0.9
746.2	Ebstein's anomaly	16	0.6	0.7	0.6	1.2	0.7	0.4	0.9
746.3	Congenital stenosis of aortic valve	36	1.4	1.7	1.1	1.5	1.9	0.2	0.5
746.7	Hypoplastic left heart syndrome	55	2.1	2.5	1.7	1.4	2.3	1.8	0.9
746.85	Anomalies of coronary artery	6	0.2	0.3	0.2	1.9	0.3	0.2	0.0
747.0	Patent ductus arteriosis	318	12.3	12.1	12.5	1.0	11.1	17.1	10.6
747.10	Coartation of aorta	110	4.3	4.8	3.7	1.3	4.6	2.9	5.1
747.41	Total anomalous pulmonary venus connect.	10	0.4	0.3	0.5	0.6	0.4	0.4	0.5
748.0	Choanal atresia	34	1.3	1.4	1.3	1.1	1.4	1.1	0.5
748.5	Agenesis/hypoplasia of lung	72	2.8	3.2	2.4	1.3	2.4	4.2	2.3
749.0	Cleft palate	145	5.6	5.2	6.0	0.9	6.5	3.1	4.2

2000 Births – New York State Residents (continued)

ICD-9 Code	Malformation	Total Number	Total Prevalence	Male	Female	Ratio (M/F)	White	Black	Other
749.1	Cleft lip	68	2.6	3.6	1.6	2.3	2.6	2.6	2.8
749.2	Cleft palate & lip	117	4.5	5.6	3.4	1.6	5.3	1.7	5.5
750.3	Tracheoesophageal fistula etc.	57	2.2	2.5	1.9	1.3	2.8	1.1	0.0
750.5	Congenital hypertrophic pyloric stenosis	394	15.2	24.3	5.7	4.2	18.0	9.7	6.0
751.1	Atresia and stenosis of small intestine	73	2.8	2.3	3.4	0.7	2.7	2.9	2.8
751.2	Atresia and stenosis of rectum or anus	84	3.2	3.7	2.8	1.3	3.3	2.9	3.2
751.3	Hirschsprungs disease	44	1.7	2.0	1.4	1.4	1.5	2.8	0.0
751.4	Anomalies of intestinal fixation	33	1.3	2.0	0.6	3.5	1.3	1.5	0.9
751.61	Biliary atresia	24	0.9	1.1	0.8	1.3	0.7	1.5	1.8
752.6	Epispadias	9	0.3	0.7	0.0		0.4	0.2	0.0
752.6	Hypospadias	893	34.5	66.9	0.4	168.6	35.6	34.3	24.9
753.0	Renal agenesis and dysgenesis	75	2.9	4.2	1.5	2.8	3.3	2.2	0.9
753.1	Cystic kidney disease	122	4.7	4.8	4.6	1.0	4.7	5.0	4.2
753.2	Obstructive defect renal pelvis & ureter	683	26.4	36.4	15.9	2.3	29.1	16.5	28.1
753.5	Extrophy of urinary bladder	6	0.2	0.2	0.2	0.9	0.3	0.0	0.0
753.6	Atresia & stenosis of urethra & bladder	13	0.5	0.8	0.2	5.2	0.4	1.0	0.0
754.3	Congenital dislocation of hip	219	8.6	3.3	14.2	0.2	10.1	4.4	6.2
754.51	Talipes equinovarus	272	10.7	12.6	8.8	1.4	11.0	9.8	10.0
755.2	Reduction deformities of upper limb	20	0.8	0.7	0.9	0.8	0.8	0.8	1.0
755.3	Reduction deformities of lower limb	33	1.3	1.5	1.1	1.5	1.3	1.0	1.9
755.8	Arthrogryposis multiplex congenita	10	0.4	0.4	0.4	1.0	0.4	0.4	0.5
756.0	Craniosynostosis	94	3.7	4.2	3.2	1.3	4.2	2.9	1.4
756.0	Goldenhar syndrome	5	0.2	0.1	0.3	0.2	0.3	0.0	0.0
756.4	Chonodrodystrophy	35	1.4	1.6	1.1	1.4	1.2	2.1	1.0
756.51	Osteogenesis imperfecta	8	0.3	0.2	0.5	0.3	0.3	0.2	0.5
756.6	Diaphragmatic hernia	28	1.1	1.5	0.7	2.0	1.1	1.4	0.5
756.7	Gastroschisis	39	1.5	1.6	1.5	1.1	1.8	1.4	0.0
756.7	Omphalocele	31	1.2	1.4	1.1	1.3	1.2	1.4	1.0
756.7	Prune belly	5	0.2	0.4	0.0		0.2	0.2	0.0
758.0	Down syndrome	285	11.2	11.3	11.2	1.0	11.9	8.3	11.9
758.1	Patau syndrome	14	0.6	0.6	0.5	1.3	0.5	0.8	0.5
758.2	Edwards syndrome	23	0.9	0.5	1.4	0.3	0.6	1.9	1.0
758.6	Gonadal dysgenesis	13	0.5	0.2	0.9	0.2	0.6	0.2	0.5
758.7	Klinefelter syndrome	14	0.6	1.1	0.0		0.6	0.4	0.5
759.3	Situs inversus	11	0.4	0.5	0.3	1.7	0.4	0.4	0.5
760.71	Fetal alcohol syndrome	15	0.6	0.8	0.3	2.6	0.2	1.9	0.0
771.0	Congenital rubella	1	0.0	0.1	0.0		0.1	0.0	0.0
771.1	Congenital cytomegalovirus infection	19	0.7	0.6	0.9	0.7	0.6	1.7	0.0
771.2	Other congenital infections	21	0.8	0.8	0.8	1.0	0.6	1.5	1.0

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

2001 Births– New York State Residents

ICD-9 Code	Malformation	Total Number	Total Prevalence	Male	Female	Ratio (M/F)	White	Black	Other/ Unknown
243	Congenital hypothyroidism	59	2.3	2.9	1.8	1.6	1.8	4.6	0.5
270.1	Phenylketonuria	2	0.1	0.2	0.0		0.1	0.0	0.0
277.0	Cystic fibrosis	20	0.8	0.6	1.0	0.6	0.9	0.4	0.5
282.6	Sickle-cell anemia	76	3.0	2.9	3.2	0.9	0.3	13.5	0.0
740.0	Anencephalus	11	0.4	0.5	0.4	1.1	0.6	0.2	0.0
741.0	Spina bifida with hydrocephalus	24	0.9	1.2	0.7	1.6	0.9	1.4	0.0
741.9	Spina bifida without hydrocephalus	31	1.2	1.0	1.5	0.7	1.2	1.4	1.0
742.0	Encephalocele	16	0.6	0.6	0.6	1.0	0.6	0.6	1.0
742.1	Microcephalus	90	3.6	3.3	3.8	0.9	3.2	5.0	3.3
742.2	Agyria & lissencephaly	6	0.2	0.2	0.2	1.0	0.3	0.0	0.0
742.2	Anomalies of corpus callosum	25	1.0	1.2	0.8	1.4	1.0	0.6	1.9
742.2	Holoprosencephaly	5	0.2	0.2	0.2	0.6	0.2	0.0	0.0
742.3	Congenital hydrocephalus	159	6.3	7.2	5.3	1.4	6.3	7.5	2.4
742.4	Porencephaly	13	0.5	0.6	0.4	1.5	0.6	0.4	0.0
742.5	Congenital tethered cord	22	0.9	0.8	0.9	1.0	0.8	0.6	2.4
743.0	Anophthalmos	4	0.2	0.1	0.2	0.3	0.2	0.0	0.5
743.1	Microphthalmos	10	0.4	0.5	0.3	1.4	0.3	0.8	0.0
743.2	Glaucoma	14	0.6	0.7	0.4	1.7	0.6	0.6	0.0
743.3	Absence of lens	7	0.3	0.2	0.3	0.7	0.3	0.2	0.5
743.3	Congenital cataract	23	0.9	0.8	1.0	0.9	0.9	1.0	0.5
743.45	Aniridia	2	0.1	0.2	0.0		0.1	0.0	0.0
743.46	Coloboma of iris	3	0.1	0.1	0.2	0.5	0.2	0.0	0.0
744.0	Anotia/microtia	6	0.2	0.2	0.2	1.0	0.3	0.0	0.5
745.0	Common truncus	11	0.4	0.4	0.5	0.8	0.4	0.4	0.5
745.1	Transposition of great vessels	91	3.6	4.4	2.7	1.6	3.5	3.1	5.7
745.2	Tetralogy of Fallot	112	4.4	4.9	4.0	1.2	4.2	4.1	7.1
745.3	Common ventricle	12	0.5	0.4	0.6	0.7	0.5	0.6	0.0
745.4	Ventricular septal defect	839	33.1	30.6	35.7	0.9	34.2	29.0	33.3
745.5	Ostium secundum type atrial septal def.	1,126	44.4	47.8	40.9	1.2	39.9	62.9	36.6
745.6	Endocardial cushion defects	42	1.7	1.2	2.1	0.6	1.6	2.5	0.5
746.0	Atresia/stenosis of pulmonary valve	211	8.3	7.7	9.0	0.9	8.0	10.2	5.7
746.1	Tricuspid atresia/stenosis/hypoplasia	10	0.4	0.4	0.4	1.0	0.4	0.6	0.0
746.2	Ebstein's anomaly	14	0.6	0.5	0.6	0.7	0.6	0.6	0.5
746.3	Congenital stenosis of aortic valve	28	1.1	1.7	0.5	3.5	1.4	0.4	0.5
746.7	Hypoplastic left heart syndrome	47	1.9	2.2	1.5	1.5	2.0	1.9	0.5
746.85	Anomalies of coronary artery	7	0.3	0.5	0.0		0.2	0.6	0.0
747.0	Patent ductus arteriosis	355	14.0	15.0	13.0	1.1	12.9	20.7	7.1
747.10	Coartation of aorta	89	3.5	4.0	3.0	1.3	3.4	3.1	5.7
747.41	Total anomalous pulmonary venus connect.	11	0.4	0.5	0.3	1.7	0.4	0.6	0.5

2001 Births – New York State Residents (continued)

ICD-9 Code	Malformation	Total Number	Total Prevalence	Male	Female	Ratio (M/F)	White	Black	Other
748.0	Choanal atresia	27	1.1	1.2	0.9	1.4	1.5	0.0	0.0
748.5	Agenesis/hypoplasia of lung	56	2.2	2.8	1.6	1.7	2.3	1.9	1.4
749.0	Cleft palate	169	6.7	6.6	6.8	1.0	8.0	3.7	2.9
749.1	Cleft lip	54	2.1	2.5	1.8	1.4	2.4	1.0	2.4
749.2	Cleft palate & lip	111	4.4	5.6	3.2	1.8	4.7	3.5	3.3
750.3	Tracheoesophageal fistula etc.	53	2.1	2.4	1.8	1.3	2.5	1.2	1.0
750.5	Congenital hypertrophic pyloric stenosis	386	15.2	24.9	5.1	4.9	17.4	8.9	11.4
751.1	Atresia and stenosis of small intestine	84	3.3	3.3	3.3	1.0	3.2	4.1	2.9
751.2	Atresia and stenosis of rectum or anus	94	3.7	4.2	3.2	1.3	3.8	2.7	5.2
751.3	Hirschsprungs disease	51	2.0	2.9	1.1	2.8	2.0	2.1	1.9
751.4	Anomalies of intestinal fixation	44	1.7	1.9	1.6	1.1	2.0	0.8	1.9
751.61	Biliary atresia	20	0.8	0.5	1.1	0.5	0.7	1.0	1.0
752.6	Epispadias	7	0.3	0.5	0.0		0.3	0.0	0.5
752.6	Hypospadias	886	35.0	67.9	0.4	168.0	37.0	31.8	23.3
753.0	Renal agenesis and dysgenesis	66	2.6	3.1	2.1	1.5	2.7	2.7	1.4
753.1	Cystic kidney disease	116	4.6	5.2	4.0	1.3	4.4	6.2	1.9
753.2	Obstructive defect renal pelvis & ureter	698	27.5	39.0	15.5	2.5	29.5	21.0	25.2
753.5	Extrophy of urinary bladder	4	0.2	0.2	0.1	2.9	0.2	0.0	0.0
753.6	Atresia & stenosis of urethra & bladder	13	0.5	0.8	0.2	5.2	0.4	1.0	0.0
754.3	Congenital dislocation of hip	219	8.6	3.3	14.2	0.2	10.1	4.4	6.2
754.51	Talipes equinovarus	272	10.7	12.6	8.8	1.4	11.0	9.8	10.0
755.2	Reduction deformities of upper limb	20	0.8	0.7	0.9	0.8	0.8	0.8	1.0
755.3	Reduction deformities of lower limb	33	1.3	1.5	1.1	1.5	1.3	1.0	1.9
755.8	Arthrogryposis multiplex congenita	10	0.4	0.4	0.4	1.0	0.4	0.4	0.5
756.0	Craniosynostosis	94	3.7	4.2	3.2	1.3	4.2	2.9	1.4
756.0	Goldenhar syndrome	5	0.2	0.1	0.3	0.2	0.3	0.0	0.0
756.4	Chonodrodystrophy	35	1.4	1.6	1.1	1.4	1.2	2.1	1.0
756.51	Osteogenesis imperfecta	8	0.3	0.2	0.5	0.3	0.3	0.2	0.5
756.6	Diaphragmatic hernia	28	1.1	1.5	0.7	2.0	1.1	1.4	0.5
756.7	Gastroschisis	39	1.5	1.6	1.5	1.1	1.8	1.4	0.0
756.7	Omphalocele	31	1.2	1.4	1.1	1.3	1.2	1.4	1.0
756.7	Prune belly	5	0.2	0.4	0.0		0.2	0.2	0.0
758.0	Down syndrome	285	11.2	11.3	11.2	1.0	11.9	8.3	11.9
758.1	Patau syndrome	14	0.6	0.6	0.5	1.3	0.5	0.8	0.5
758.2	Edwards syndrome	23	0.9	0.5	1.4	0.3	0.6	1.9	1.0
758.6	Gonadal dysgenesis	13	0.5	0.2	0.9	0.2	0.6	0.2	0.5
758.7	Klinefelter syndrome	14	0.6	1.1	0.0		0.6	0.4	0.5
759.3	Situs inversus	11	0.4	0.5	0.3	1.7	0.4	0.4	0.5
760.71	Fetal alcohol syndrome	15	0.6	0.8	0.3	2.6	0.2	1.9	0.0
771.0	Congenital rubella	1	0.0	0.1	0.0		0.1	0.0	0.0
771.1	Congenital cytomegalovirus infection	19	0.7	0.6	0.9	0.7	0.6	1.7	0.0
771.2	Other congenital infections	21	0.8	0.8	0.8	1.0	0.6	1.5	1.0

Section IV
Most Frequently Reported Selected Major Malformations by County

Introduction to Tables

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

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

Section IV. Children with Major Congenital Malformations & Percent of Live Births by County and Birth Year, 1998-2001

	1998		1999		2000		2001	
County	Number of Children	Percent of Live Births	Number of Children	Percent of Live Births	Number of Children	Percent of Live Births	Number of Children	Percent of Live Births
Albany	177	5.1	133	4.2	122	3.7	147	4.6
Allegany	24	4.4	28	5.0	19	3.5	12	2.3
Bronx	978	4.3	822	3.7	859	3.8	835	3.6
Broome	96	4.3	102	4.5	95	4.3	101	4.7
Cattaraugus	48	4.5	42	4.2	52	5.1	39	3.9
Cayuga	41	4.5	43	4.7	38	4.0	47	5.3
Chautauqua	65	4.2	58	3.8	65	4.2	66	4.2
Chemung	48	4.7	42	3.8	50	4.6	51	4.7
Chenango	24	4.0	21	3.9	36	6.3	33	5.7
Clinton	16	2.2	19	2.3	35	4.5	19	2.7
Columbia	18	3.0	17	2.9	24	3.6	23	3.9
Cortland	37	6.1	30	5.2	28	5.0	21	3.8
Delaware	17	3.7	14	3.1	16	3.5	15	3.6
Dutchess	106	3.2	127	4.0	124	3.7	132	4.1
Erie	655	5.7	612	5.5	655	5.8	631	5.8
Essex	7	1.9	9	2.3	7	2.0	5	1.5
Franklin	15	3.1	13	2.9	13	2.8	12	2.7
Fulton	43	7.0	29	5.0	41	6.8	49	7.9
Genesee	31	4.5	40	5.4	44	6.4	29	4.1
Greene	12	2.4	23	5.3	16	3.3	16	3.9
Hamilton	1	2.6	1	3.2	5	10.4	2	3.8
Herkimer	26	3.9	22	3.3	31	4.7	28	4.2
Jefferson	57	3.3	66	3.8	112	6.8	81	4.7
Kings	1874	4.7	1754	4.4	1760	4.4	1762	4.4
Lewis	15	4.4	11	3.3	17	5.1	25	7.3
Livingston	36	5.4	35	5.0	22	3.3	30	4.7
Madison	34	3.9	40	5.2	36	4.6	38	5.0
Monroe	395	4.1	369	4.0	360	3.8	338	3.8
Montgomery	21	3.5	19	3.3	18	2.9	25	4.1
Nassau	887	5.2	757	4.5	771	4.5	826	5.1
New York	699	3.6	622	3.2	648	3.2	604	3.0
Niagara	132	5.0	120	4.6	145	5.8	111	4.5
Oneida	123	4.7	130	4.9	140	5.6	125	5.0
Onondaga	279	4.7	290	4.8	277	4.6	291	5.0
Ontario	58	4.9	46	4.3	48	4.2	36	3.2
Orange	173	3.6	161	3.3	168	3.4	165	3.3
Orleans	23	4.2	23	4.3	33	6.2	24	5.2
Oswego	57	3.9	71	4.8	67	4.5	66	4.6
Otsego	20	3.5	13	2.3	15	2.6	20	3.8
Putnam	40	3.2	45	3.8	52	4.4	44	3.7
Queens	1197	3.7	1256	4.0	1275	4.0	1169	3.8
Rensselear	66	3.7	70	4.0	89	5.2	69	4.1
Richmond	251	4.4	264	4.5	249	4.2	172	3.1
Rockland	170	3.8	137	3.0	161	3.5	166	3.6
Saratoga	87	3.4	82	3.4	108	4.3	109	4.5
Schenectady	60	3.5	72	4.2	62	3.6	72	4.2

Section IV. Children with Major Congenital Malformations & Percent of Live Births by County and Birth Year, 1998-2001 (continued)

	1998		1999		2000		2001	
County	Number of Children	Percent of Live Births	Number of Children	Percent of Live Births	Number of Children	Percent of Live Births	Number of Children	Percent of Live Births
Schoharie	8	2.5	17	5.5	7	2.2	13	4.3
Schuyler	9	4.2	8	3.7	7	3.2	8	3.7
Seneca	14	3.8	17	4.7	8	2.2	8	2.2
St Lawrence	43	3.6	47	4.1	35	2.9	62	5.0
Steuben	42	3.6	43	3.7	76	6.4	59	5.0
Suffolk	768	3.9	780	3.9	760	3.8	753	3.8
Sullivan	27	3.3	43	5.0	31	3.7	23	2.7
Tioga	20	3.6	18	2.9	15	2.5	19	3.2
Tompkins	31	3.7	33	3.7	49	5.9	32	3.7
Ulster	74	4.0	79	4.5	79	4.4	83	4.6
Warren	15	2.3	23	3.5	35	5.6	25	3.7
Washington	24	4.1	18	2.8	20	3.3	22	3.8
Wayne	46	3.6	43	3.5	37	3.2	38	3.5
Westchester	502	3.9	441	3.5	514	3.9	440	3.4
Wyoming	30	6.5	22	5.2	25	5.8	17	3.9
Yates	10	2.9	10	3.1	14	5.1	11	3.6

Section IV
Most Frequently Reported Major Congenital Malformations by County, 1998-2001
Congenital Malformations Registry

County	ICD-9 Code	Description	Number in 1998-2001
Albany	745.5	Ostium secundum atrial septal defect	72
	745.4	Ventricular septal defect	56
	752.5	Undescended testicle	56
	755	Polydactyly	47
	752.6	Hypospadias & epispadias	46
	754.5	Varus deformities of feet	24
	754.3	Congenital dislocation of hip	23
	747	Patent ductus arteriosus	22
	750.5	Congenital hypertrophic pyloric stenosis	20
	753.2	Obstructive defects of renal pelvis & ureter	18
Allegany	745.5	Ostium secundum atrial septal defect	13
	752.5	Undescended testicle	10
	747.3	Anomalies of pulmonary artery	9
	746.8	Other specified anomalies of heart	8
	750.5	Congenital hypertrophic pyloric stenosis	6
	745.4	Ventricular septal defect	5
	753.2	Obstructive defects of renal pelvis & ureter	5
	752.6	Hypospadias & epispadias	4
	755	Polydactyly	4
	748.3	Other anomalies of larynx, trachea, & bronchus	3
	754.3	Congenital dislocation of hip	3
	754.5	Varus deformities of feet	3
	756	Anomalies of skull and face bones	3
	758	Down syndrome	3
	759.8	Other specified anomalies	3
Bronx	755	Polydactyly	305
	752.6	Hypospadias & epispadias	302
	745.4	Ventricular septal defect	276
	752.5	Undescended testicle	264
	745.5	Ostium secundum atrial septal defect	263
	753.2	Obstructive defects of renal pelvis & ureter	230
	754.5	Varus deformities of feet	160
	750.5	Congenital hypertrophic pyloric stenosis	146
	754.3	Congenital dislocation of hip	93
Broome	745.5	Ostium secundum atrial septal defect	44
	745.4	Ventricular septal defect	39
	752.6	Hypospadias & epispadias	32
	754.5	Varus deformities of feet	26
	753.2	Obstructive defects of renal pelvis & ureter	24
	746.0	Anomalies of pulmonary valve	23

Section IV
Most Frequently Reported Major Congenital Malformations by County, 1998-2001
Congenital Malformations Registry

County	ICD-9 Code	Description	Number in 1998-2001
Broome	752.5	Undescended testicle	23
	755.0	Polydactyly	17
	750.5	Congenital hypertrophic pyloric stenosis	14
	747.0	Patent ductus arteriosus	13
	758.0	Down syndrome	13
Cattarugus	745.5	Ostium secundum atrial septal defect	28
	745.4	Ventricular septal defect	21
	754.5	Varus deformities of feet	17
	752.5	Undescended testicle	12
	752.6	Hypospadias & epispadias	12
	747.3	Anomalies of pulmonary artery	11
	755.6	Other anomalies of lower limb including pelvic girdle	11
	746.8	Other specified anomalies of heart	9
	750.5	Congenital hypertrophic pyloric stenosis	7
	742.3	Congenital hydrocephalus	6
	750.3	Tracheoesophageal fistula, esophageal atresia & stenosis	6
	751.2	Atresia & stenosis of large intestine, rectum, & anal canal	6
	758.0	Down syndrome	6
Cayuga	752.6	Hypospadias & epispadias	17
	745.4	Ventricular septal defect	16
	745.5	Ostium secundum atrial septal defect	15
	753.2	Obstructive defects of renal pelvis & ureter	11
	750.5	Congenital hypertrophic pyloric stenosis	10
	752.5	Undescended testicle	10
	758.0	Down syndrome	10
	747.0	Patent ductus arteriosus	8
	754.5	Varus deformities of feet	8
	524.0	Major anomalies of jaw size	7
	748.3	Other anomalies of larynx, trachea, & bronchus	7
	755.0	Polydactyly	7
	756.0	Anomalies of skull and face bones	7
Chautauqua	745.5	Ostium secundum atrial septal defect	61
	752.6	Hypospadias & epispadias	30
	745.4	Ventricular septal defect	23
	746.8	Other specified anomalies of heart	20
	752.5	Undescended testicle	19
	747.3	Anomalies of pulmonary artery	17
	754.5	Varus deformities of feet	13
	755.0	Polydactyly	13
	748.3	Other anomalies of larynx, trachea, & bronchus	10
	753.2	Obstructive defects of renal pelvis & ureter	8
	756.0	Anomalies of skull and face bones	8

Section IV
Most Frequently Reported Major Congenital Malformations by County, 1998-2001
Congenital Malformations Registry

County	ICD-9 Code	Description	Number in 1998-2001
Chemung	745.4	Ventricular septal defect	24
	752.5	Undescended testicle	20
	752.6	Hypospadias & epispadias	18
	753.2	Obstructive defects of renal pelvis & ureter	18
	750.5	Congenital hypertrophic pyloric stenosis	12
	745.5	Ostium secundum atrial septal defect	8
	754.3	Congenital dislocation of hip	8
	751.5	Other anomalies of intestine	7
	754.5	Varus deformities of feet	6
	745.2	Tetralogy of Fallot	5
	746.8	Other specified anomalies of heart	5
Chenango	745.5	Ostium secundum atrial septal defect	18
	750.5	Congenital hypertrophic pyloric stenosis	15
	752.6	Hypospadias & epispadias	12
	745.4	Ventricular septal defect	8
	746.0	Anomalies of pulmonary valve	7
	754.5	Varus deformities of feet	7
	747.3	Anomalies of pulmonary artery	6
	742.3	Congenital hydrocephalus	5
	758.0	Down syndrome	5
	755.1	Syndactyly	4
Clinton	745.5	Ostium secundum atrial septal defect	13
	752.6	Hypospadias & epispadias	13
	752.5	Undescended testicle	10
	753.2	Obstructive defects of renal pelvis & ureter	8
	745.4	Ventricular septal defect	7
	747.0	Patent ductus arteriosus	5
	752.8	Other specified anomalies of genital organs	5
	758.0	Down syndrome	4
	746.8	Other specified anomalies of heart	3
	754.3	Congenital dislocation of hip	3
Columbia	745.5	Ostium secundum atrial septal defect	15
	752.6	Hypospadias & epispadias	10
	752.5	Undescended testicle	8
	746.8	Other specified anomalies of heart	7
	754.3	Congenital dislocation of hip	6
	745.4	Ventricular septal defect	5
	747.0	Patent ductus arteriosus	5
	750.5	Congenital hypertrophic pyloric stenosis	5
	751.1	Atresia & stenosis of small intestine	4
	756.7	Anomalies of abdominal wall	4

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County	ICD-9 Code	Description	Number in 1998-2001
Cortland	752.6	Hypospadias & epispadias	12
	745.4	Ventricular septal defect	11
	745.5	Ostium secundum atrial septal defect	10
	753.2	Obstructive defects of renal pelvis & ureter	8
	750.5	Congenital hypertrophic pyloric stenosis	6
	754.5	Varus deformities of feet	6
	756.0	Anomalies of skull and face bones	6
	749.0	Cleft palate	5
	746.0	Anomalies of pulmonary valve	4
	749.1	Cleft lip	4
	749.2	Cleft palate with cleft lip	4
	752.5	Undescended testicle	4
	754.6	Valgus deformities of feet	4
	755.0	Polydactyly	4
Delaware	745.5	Ostium secundum atrial septal defect	7
	752.6	Hypospadias & epispadias	6
	750.5	Congenital hypertrophic pyloric stenosis	5
	752.5	Undescended testicle	5
	745.1	Transposition of great vessels	3
	745.4	Ventricular septal defect	3
	746.8	Other specified anomalies of heart	3
	747.1	Coarctation of aorta	3
	747.3	Anomalies of pulmonary artery	3
	753.2	Obstructive defects of renal pelvis & ureter	3
	756.0	Anomalies of skull and face bones	3
Dutchess	752.6	Hypospadias & epispadias	55
	752.5	Undescended testicle	49
	745.4	Ventricular septal defect	36
	753.2	Obstructive defects of renal pelvis & ureter	30
	750.5	Congenital hypertrophic pyloric stenosis	25
	754.3	Congenital dislocation of hip	23
	755.0	Polydactyly	22
	754.5	Varus deformities of feet	20
	745.5	Ostium secundum atrial septal defect	17
	746.0	Anomalies of pulmonary valve	17
	755.6	Other anomalies of lower limb including pelvic girdle	17
Erie	745.5	Ostium secundum atrial septal defect	537
	747.3	Anomalies of pulmonary artery	240
	752.6	Hypospadias & epispadias	234

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County	ICD-9 Code	Description	Number in 1998-2001
Erie	752.5	Undescended testicle	221
	745.4	Ventricular septal defect	217
	746.8	Other specified anomalies of heart	205
	755.0	Polydactyly	145
	750.5	Congenital hypertrophic pyloric stenosis	103
	748.3	Other anomalies of larynx, trachea, & bronchus	91
	754.5	Varus deformities of feet	79
Essex	745.5	Ostium secundum atrial septal defect	4
	752.5	Undescended testicle	4
	752.6	Hypospadias & epispadias	3
	745.4	Ventricular septal defect	2
	756.0	Anomalies of skull and face bones	2
	228.0	Hemangioma, any site	1
	742.3	Congenital hydrocephalus	1
	742.4	Other specified anomalies of brain	1
	743.3	Congenital cataract & lens anomalies	1
	747.0	Patent ductus arteriosus	1
	748.5	Agenesis, hypoplasia & dysplasia, lung	1
	749.1	Cleft lip	1
	750.5	Congenital hypertrophic pyloric stenosis	1
	751.1	Atresia & stenosis of small intestine	1
	751.6	Anomalies of gallbladder, bile ducts, and liver	1
	752.8	Other specified anomalies of genital organs	1
	753.1	Cystic kidney disease	1
	753.2	Obstructive defects of renal pelvis & ureter	1
	753.8	Other specified anomalies of bladder & urethra	1
	757.6	Specified anomalies of breast	1
	758.8	Other conditions due to sex chromosome anomalies	1
Franklin	752.6	Hypospadias & epispadias	15
	753.2	Obstructive defects of renal pelvis & ureter	5
	749.0	Cleft palate	4
	745.4	Ventricular septal defect	3
	752.5	Undescended testicle	3
	758.0	Down syndrome	3
	270.6	Disorders of urea cycle metablism	2
	745.5	Ostium secundum atrial septal defect	2
	747.1	Coarctation of aorta	2
	748.3	Other anomalies of larynx, trachea, & bronchus	2
	749.2	Cleft palate with cleft lip	2
	752.8	Other specified anomalies of genital organs	2
	754.3	Congenital dislocation of hip	2

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County	ICD-9 Code	Description	Number in 1998-2001
Fulton	752.6	Hypospadias & epispadias	17
	745.4	Ventricular septal defect	14
	745.5	Ostium secundum atrial septal defect	12
	746.8	Other specified anomalies of heart	11
	754.3	Congenital dislocation of hip	10
	754.5	Varus deformities of feet	10
	750.5	Congenital hypertrophic pyloric stenosis	9
	752.5	Undescended testicle	9
	754.6	Valgus deformities of feet	6
Genesee	745.5	Ostium secundum atrial septal defect	26
	752.6	Hypospadias & epispadias	19
	745.4	Ventricular septal defect	15
	752.5	Undescended testicle	13
	754.3	Congenital dislocation of hip	11
	746.8	Other specified anomalies of heart	10
	753.2	Obstructive defects of renal pelvis & ureter	9
	750.5	Congenital hypertrophic pyloric stenosis	7
	748.3	Other anomalies of larynx, trachea, & bronchus	6
	752.8	Other specified anomalies of genital organs	5
	755.1	Syndactyly	5
Greene	745.5	Ostium secundum atrial septal defect	13
	752.6	Hypospadias & epispadias	13
	745.4	Ventricular septal defect	11
	742.2	Reduction deformities of brain	4
	756.0	Anomalies of skull and face bones	4
	742.3	Congenital hydrocephalus	3
	746.0	Anomalies of pulmonary valve	3
	750.3	Tracheoesophageal fistula, esophageal atresia & stenosis	3
	753.2	Obstructive defects of renal pelvis & ureter	3
	754.3	Congenital dislocation of hip	3
Hamilton	745.5	Ostium secundum atrial septal defect	2
	755.0	Polydactyly	2
	758.0	Down syndrome	2
	745.4	Ventricular septal defect	1
	746.0	Anomalies of pulmonary valve	1
	746.8	Other specified anomalies of heart	1
	748.3	Other anomalies of larynx, trachea, & bronchus	1
	753.2	Obstructive defects of renal pelvis & ureter	1
	756.7	Anomalies of abdominal wall	1

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County	ICD-9 Code	Description	Number in 1998-2001
Herkimer	745.4	Ventricular septal defect	10
	745.5	Ostium secundum atrial septal defect	10
	752.5	Undescended testicle	8
	750.5	Congenital hypertrophic pyloric stenosis	7
	753.2	Obstructive defects of renal pelvis & ureter	7
	754.3	Congenital dislocation of hip	7
	746.8	Other specified anomalies of heart	6
	756.0	Anomalies of skull and face bones	6
	758.0	Down syndrome	6
	742.4	Other specified anomalies of brain	5
	746.7	Hypoplastic left heart syndrome	5
Jefferson	745.4	Ventricular septal defect	32
	752.6	Hypospadias & epispadias	32
	752.5	Undescended testicle	28
	753.2	Obstructive defects of renal pelvis & ureter	27
	755.0	Polydactyly	17
	750.5	Congenital hypertrophic pyloric stenosis	14
	745.5	Ostium secundum atrial septal defect	13
	748.3	Other anomalies of larynx, trachea, & bronchus	12
	742.3	Congenital hydrocephalus	11
	754.5	Varus deformities of feet	11
Kings	745.5	Ostium secundum atrial septal defect	1413
	745.4	Ventricular septal defect	582
	752.6	Hypospadias & epispadias	538
	755.0	Polydactyly	456
	752.5	Undescended testicle	369
	753.2	Obstructive defects of renal pelvis & ureter	363
	747.0	Patent ductus arteriosus	251
	747.3	Anomalies of pulmonary artery	210
	750.5	Congenital hypertrophic pyloric stenosis	210
	754.5	Varus deformities of feet	207
Lewis	754.3	Congenital dislocation of hip	8
	758.0	Down syndrome	8
	745.4	Ventricular septal defect	6
	745.5	Ostium secundum atrial septal defect	6
	752.6	Hypospadias & epispadias	6
	749.0	Cleft palate	4
	754.5	Varus deformities of feet	4
	756.0	Anomalies of skull and face bones	4
	243.0	Congenital hypothyroidism	3

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County	ICD-9 Code	Description	Number in 1998-2001
Lewis	746.8	Other specified anomalies of heart	3
	752.5	Undescended testicle	3
Livingston	752.6	Hypospadias & epispadias	15
	753.2	Obstructive defects of renal pelvis & ureter	15
	745.4	Ventricular septal defect	11
	745.5	Ostium secundum atrial septal defect	10
	752.5	Undescended testicle	10
	750.5	Congenital hypertrophic pyloric stenosis	7
	755.1	Syndactyly	5
	755.6	Other anomalies of lower limb including pelvic girdle	5
	749.1	Cleft lip	4
	753.0	Renal agenesis & dysgenesis	4
	754.5	Varus deformities of feet	4
	758.0	Down syndrome	4
Madison	752.5	Undescended testicle	16
	752.6	Hypospadias & epispadias	12
	745.4	Ventricular septal defect	11
	754.3	Congenital dislocation of hip	11
	745.5	Ostium secundum atrial septal defect	9
	750.5	Congenital hypertrophic pyloric stenosis	8
	748.3	Other anomalies of larynx, trachea, & bronchus	7
	755.0	Polydactyly	7
	753.2	Obstructive defects of renal pelvis & ureter	6
	756.0	Anomalies of skull and face bones	6
Monroe	752.6	Hypospadias & epispadias	188
	753.2	Obstructive defects of renal pelvis & ureter	167
	745.4	Ventricular septal defect	122
	752.5	Undescended testicle	114
	755.0	Polydactyly	93
	754.5	Varus deformities of feet	65
	745.5	Ostium secundum atrial septal defect	63
	750.5	Congenital hypertrophic pyloric stenosis	51
	758.0	Down syndrome	45
	746.8	Other specified anomalies of heart	44
Montgomery	752.6	Hypospadias & epispadias	11
	752.5	Undescended testicle	8
	745.4	Ventricular septal defect	6
	746.8	Other specified anomalies of heart	6
	753.2	Obstructive defects of renal pelvis & ureter	5

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County	ICD-9 Code	Description	Number in 1998-2001
Montgomery	754.5	Varus deformities of feet	5
	745.5	Ostium secundum atrial septal defect	4
	749.0	Cleft palate	4
	749.2	Cleft palate with cleft lip	4
	755.0	Polydactyly	4
Nassau	752.6	Hypospadias & epispadias	394
	745.5	Ostium secundum atrial septal defect	364
	752.5	Undescended testicle	295
	745.4	Ventricular septal defect	287
	753.2	Obstructive defects of renal pelvis & ureter	232
	750.5	Congenital hypertrophic pyloric stenosis	138
	754.3	Congenital dislocation of hip	118
	755.0	Polydactyly	106
	747.0	Patent ductus arteriosus	102
	746.8	Other specified anomalies of heart	101
	754.5	Varus deformities of feet	101
New York	745.5	Ostium secundum atrial septal defect	288
	745.4	Ventricular septal defect	262
	752.6	Hypospadias & epispadias	226
	753.2	Obstructive defects of renal pelvis & ureter	163
	752.5	Undescended testicle	153
	755.0	Polydactyly	141
	754.3	Congenital dislocation of hip	120
	750.5	Congenital hypertrophic pyloric stenosis	91
	754.5	Varus deformities of feet	69
Niagara	745.5	Ostium secundum atrial septal defect	96
	752.5	Undescended testicle	42
	745.4	Ventricular septal defect	40
	746.8	Other specified anomalies of heart	40
	750.5	Congenital hypertrophic pyloric stenosis	39
	752.6	Hypospadias & epispadias	36
	747.3	Anomalies of pulmonary artery	33
	753.2	Obstructive defects of renal pelvis & ureter	18
	755.0	Polydactyly	18
	754.5	Varus deformities of feet	15
Oneida	752.5	Undescended testicle	56
	752.6	Hypospadias & epispadias	44
	745.5	Ostium secundum atrial septal defect	34
	745.4	Ventricular septal defect	32

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County	ICD-9 Code	Description	Number in 1998-2001
Oneida	754.3	Congenital dislocation of hip	31
	750.5	Congenital hypertrophic pyloric stenosis	25
	753.2	Obstructive defects of renal pelvis & ureter	23
	754.5	Varus deformities of feet	22
	755.0	Polydactyly	20
	756.0	Anomalies of skull and face bones	19
	758.0	Down syndrome	19
Onondaga	752.6	Hypospadias & epispadias	120
	745.5	Ostium secundum atrial septal defect	108
	745.4	Ventricular septal defect	101
	752.5	Undescended testicle	74
	753.2	Obstructive defects of renal pelvis & ureter	73
	754.3	Congenital dislocation of hip	51
	750.5	Congenital hypertrophic pyloric stenosis	48
	755.0	Polydactyly	48
	746.8	Other specified anomalies of heart	36
	748.3	Other anomalies of larynx, trachea, & bronchus	36
	754.5	Varus deformities of feet	36
Ontario	752.6	Hypospadias & epispadias	37
	753.2	Obstructive defects of renal pelvis & ureter	17
	752.5	Undescended testicle	13
	754.3	Congenital dislocation of hip	13
	745.4	Ventricular septal defect	11
	750.5	Congenital hypertrophic pyloric stenosis	11
	746.8	Other specified anomalies of heart	9
	755.0	Polydactyly	8
	754.5	Varus deformities of feet	7
	758.0	Down syndrome	7
Orange	752.6	Hypospadias & epispadias	82
	745.4	Ventricular septal defect	63
	752.5	Undescended testicle	60
	753.2	Obstructive defects of renal pelvis & ureter	59
	745.5	Ostium secundum atrial septal defect	39
	750.5	Congenital hypertrophic pyloric stenosis	31
	754.3	Congenital dislocation of hip	31
	758.0	Down syndrome	26
	755.0	Polydactyly	25
	747.0	Patent ductus arteriosus	23
Orleans	745.5	Ostium secundum atrial septal defect	18

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County	ICD-9 Code	Description	Number in 1998-2001
Orleans	752.6	Hypospadias & epispadias	11
	745.4	Ventricular septal defect	9
	750.5	Congenital hypertrophic pyloric stenosis	9
	753.2	Obstructive defects of renal pelvis & ureter	8
	754.5	Varus deformities of feet	7
	752.5	Undescended testicle	6
	746.8	Other specified anomalies of heart	5
	747.3	Anomalies of pulmonary artery	4
	758.0	Down syndrome	4
Oswego	752.5	Undescended testicle	29
	752.6	Hypospadias & epispadias	29
	745.4	Ventricular septal defect	18
	745.5	Ostium secundum atrial septal defect	17
	750.5	Congenital hypertrophic pyloric stenosis	16
	753.2	Obstructive defects of renal pelvis & ureter	16
	749.0	Cleft palate	11
	754.3	Congenital dislocation of hip	11
	756.0	Anomalies of skull and face bones	10
	754.5	Varus deformities of feet	9
	755.0	Polydactyly	9
Otsego	745.5	Ostium secundum atrial septal defect	10
	752.6	Hypospadias & epispadias	8
	745.4	Ventricular septal defect	7
	758.0	Down syndrome	6
	753.2	Obstructive defects of renal pelvis & ureter	5
	752.5	Undescended testicle	4
	749.2	Cleft palate with cleft lip	3
	742.3	Congenital hydrocephalus	2
	745.2	Tetralogy of Fallot	2
	746.0	Anomalies of pulmonary valve	2
	747.0	Patent ductus arteriosus	2
	748.3	Other anomalies of larynx, trachea, & bronchus	2
	749.0	Cleft palate	2
	750.3	Tracheoesophageal fistula, esophageal atresia & stenosis	2
	751.2	Atresia & stenosis of large intestine, rectum, & anal canal	2
	752.8	Other specified anomalies of genital organs	2
	754.5	Varus deformities of feet	2
	754.6	Valgus deformities of feet	2
	755.1	Syndactyly	2
	756.0	Anomalies of skull and face bones	2
	759.8	Other specified anomalies	2

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County	ICD-9 Code	Description	Number in 1998-2001
Putnam	752.6	Hypospadias & epispadias	27
	752.5	Undescended testicle	18
	745.4	Ventricular septal defect	17
	753.2	Obstructive defects of renal pelvis & ureter	15
	745.5	Ostium secundum atrial septal defect	13
	754.3	Congenital dislocation of hip	10
	754.5	Varus deformities of feet	10
	750.5	Congenital hypertrophic pyloric stenosis	8
	742.3	Congenital hydrocephalus	7
	747.0	Patent ductus arteriosus	7
Queens	745.5	Ostium secundum atrial septal defect	523
	752.5	Undescended testicle	409
	752.6	Hypospadias & epispadias	377
	745.4	Ventricular septal defect	376
	753.2	Obstructive defects of renal pelvis & ureter	347
	755.0	Polydactyly	281
	754.3	Congenital dislocation of hip	249
	750.5	Congenital hypertrophic pyloric stenosis	216
	754.5	Varus deformities of feet	156
	747.3	Anomalies of pulmonary artery	136
Rensselaer	752.6	Hypospadias & epispadias	35
	745.5	Ostium secundum atrial septal defect	33
	752.5	Undescended testicle	30
	745.4	Ventricular septal defect	26
	754.5	Varus deformities of feet	15
	754.3	Congenital dislocation of hip	14
	749.2	Cleft palate with cleft lip	10
	753.2	Obstructive defects of renal pelvis & ureter	10
	747.3	Anomalies of pulmonary artery	9
	747.0	Patent ductus arteriosus	8
Richmond	745.5	Ostium secundum atrial septal defect	182
	752.6	Hypospadias & epispadias	100
	745.4	Ventricular septal defect	73
	752.5	Undescended testicle	64
	753.2	Obstructive defects of renal pelvis & ureter	60
	750.5	Congenital hypertrophic pyloric stenosis	55
	755.0	Polydactyly	49
	754.5	Varus deformities of feet	30
	758.0	Down syndrome	26
	746.0	Anomalies of pulmonary valve	22

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County	ICD-9 Code	Description	Number in 1998-2001
Rockland	752.6	Hypospadias & epispadias	80
	752.5	Undescended testicle	65
	745.4	Ventricular septal defect	54
	755.0	Polydactyly	37
	745.5	Ostium secundum atrial septal defect	34
	753.2	Obstructive defects of renal pelvis & ureter	29
	758.0	Down syndrome	26
	754.5	Varus deformities of feet	25
	750.5	Congenital hypertrophic pyloric stenosis	22
	754.3	Congenital dislocation of hip	17
Saratoga	752.5	Undescended testicle	58
	752.6	Hypospadias & epispadias	56
	745.5	Ostium secundum atrial septal defect	30
	750.5	Congenital hypertrophic pyloric stenosis	27
	745.4	Ventricular septal defect	23
	753.2	Obstructive defects of renal pelvis & ureter	20
	754.3	Congenital dislocation of hip	20
	747.0	Patent ductus arteriosus	15
	754.5	Varus deformities of feet	13
	755.0	Polydactyly	13
	758.0	Down syndrome	13
Schenectady	752.6	Hypospadias & epispadias	37
	752.5	Undescended testicle	28
	745.5	Ostium secundum atrial septal defect	23
	745.4	Ventricular septal defect	22
	755.0	Polydactyly	20
	753.2	Obstructive defects of renal pelvis & ureter	17
	750.5	Congenital hypertrophic pyloric stenosis	15
	754.5	Varus deformities of feet	14
	758.0	Down syndrome	12
	746.8	Other specified anomalies of heart	10
Schoharie	745.5	Ostium secundum atrial septal defect	10
	745.4	Ventricular septal defect	5
	746.8	Other specified anomalies of heart	4
	747.0	Patent ductus arteriosus	3
	750.5	Congenital hypertrophic pyloric stenosis	3
	752.5	Undescended testicle	3
	752.6	Hypospadias & epispadias	3
	753.2	Obstructive defects of renal pelvis & ureter	3
	751.2	Atresia & stenosis of large intestine, rectum, & anal canal	2

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County	ICD-9 Code	Description	Number in 1998-2001
Schoharie	753.4	Other specified anomalies of ureter	2
	754.5	Varus deformities of feet	2
	755.6	Other anomalies of lower limb including pelvic girdle	2
	758.0	Down syndrome	2
Schuyler	752.6	Hypospadias & epispadias	5
	748.4	Congenital cystic lung	2
	750.5	Congenital hypertrophic pyloric stenosis	2
	752.5	Undescended testicle	2
	755.6	Other anomalies of lower limb including pelvic girdle	2
	282.7	Other hemoglobinopathies	1
	740.0	Anencephalus	1
	742.4	Other specified anomalies of brain	1
	745.4	Ventricular septal defect	1
	745.5	Ostium secundum atrial septal defect	1
	746.7	Hypoplastic left heart syndrome	1
	747.0	Patent ductus arteriosus	1
	747.1	Coarctation of aorta	1
	747.3	Anomalies of pulmonary artery	1
	751.1	Atresia & stenosis of small intestine	1
	752.7	Indeterminate sex & pseudo-hermaphroditism	1
	753.2	Obstructive defects of renal pelvis & ureter	1
	753.3	Other specified anomalies of kidney	1
	754.0	Deformities of skull, face, & jaw	1
	754.3	Congenital dislocation of hip	1
	754.5	Varus deformities of feet	1
	755.1	Syndactyly	1
	756.0	Anomalies of skull and face bones	1
	756.8	Other specified anomalies of muscle, tendon, fascia, etc.	1
	758.0	Down syndrome	1
Seneca	752.5	Undescended testicle	6
	750.5	Congenital hypertrophic pyloric stenosis	4
	752.6	Hypospadias & epispadias	4
	755.0	Polydactyly	4
	745.4	Ventricular septal defect	3
	754.3	Congenital dislocation of hip	3
	753.2	Obstructive defects of renal pelvis & ureter	2
	228.0	Hemangioma, any site	1
	243.0	Congenital hypothyroidism	1
	255.2	Adrenogenital disorders	1
	277.0	Cystic fibrosis	1
	343.9	Infantile cerebral palsy unspecified	1

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County	ICD-9 Code	Description	Number in 1998-2001
Seneca	742.2	Reduction deformities of brain	1
	744.0	Anomalies of ear causing impairment of hearing	1
	747.1	Coarctation of aorta	1
	747.3	Anomalies of pulmonary artery	1
	747.8	Other specified anomalies of circulatory system	1
	749.0	Cleft palate	1
	749.1	Cleft lip	1
	754.0	Deformities of skull, face, & jaw	1
	754.5	Varus deformities of feet	1
	755.1	Syndactyly	1
	755.5	Other anomalies of upper limb including shoulder girdle	1
	755.6	Other anomalies of lower limb including pelvic girdle	1
	756.3	Other anomalies of ribs and sternum	1
	756.7	Anomalies of abdominal wall	1
	758.0	Down syndrome	1
St.Lawrence	754.5	Varus deformities of feet	31
	752.6	Hypospadias & epispadias	24
	745.5	Ostium secundum atrial septal defect	12
	745.4	Ventricular septal defect	11
	758.0	Down syndrome	9
	750.5	Congenital hypertrophic pyloric stenosis	8
	752.5	Undescended testicle	7
	753.2	Obstructive defects of renal pelvis & ureter	7
	746.8	Other specified anomalies of heart	6
	753.1	Cystic kidney disease	6
	754.6	Valgus deformities of feet	6
	755.0	Polydactyly	6
Steuben	745.4	Ventricular septal defect	20
	752.6	Hypospadias & epispadias	16
	752.5	Undescended testicle	15
	754.5	Varus deformities of feet	15
	745.5	Ostium secundum atrial septal defect	14
	753.2	Obstructive defects of renal pelvis & ureter	14
	747.0	Patent ductus arteriosus	13
	750.5	Congenital hypertrophic pyloric stenosis	11
	755.0	Polydactyly	8
	758.0	Down syndrome	7
Suffolk	752.6	Hypospadias & epispadias	374
	745.5	Ostium secundum atrial septal defect	360
	752.5	Undescended testicle	280

Section IV
Most Frequently Reported Major Congenital Malformations by County, 1998-2001
Congenital Malformations Registry

County	ICD-9 Code	Description	Number in 1998-2001
Suffolk	745.4	Ventricular septal defect	271
	753.2	Obstructive defects of renal pelvis & ureter	213
	750.5	Congenital hypertrophic pyloric stenosis	145
	754.3	Congenital dislocation of hip	114
	755.0	Polydactyly	110
	758.0	Down syndrome	102
	754.5	Varus deformities of feet	93
Sullivan	753.2	Obstructive defects of renal pelvis & ureter	13
	752.6	Hypospadias & epispadias	10
	754.5	Varus deformities of feet	10
	745.4	Ventricular septal defect	9
	745.5	Ostium secundum atrial septal defect	9
	752.5	Undescended testicle	9
	755.0	Polydactyly	9
	747.3	Anomalies of pulmonary artery	7
	750.5	Congenital hypertrophic pyloric stenosis	6
	746.8	Other specified anomalies of heart	4
	747.1	Coarctation of aorta	4
Tioga	745.5	Ostium secundum atrial septal defect	15
	753.2	Obstructive defects of renal pelvis & ureter	9
	745.4	Ventricular septal defect	8
	752.6	Hypospadias & epispadias	7
	747.3	Anomalies of pulmonary artery	5
	750.5	Congenital hypertrophic pyloric stenosis	4
	752.5	Undescended testicle	4
	742.3	Congenital hydrocephalus	3
	746.8	Other specified anomalies of heart	3
	749.0	Cleft palate	3
	749.2	Cleft palate with cleft lip	3
	758.0	Down syndrome	3
Tompkins	745.4	Ventricular septal defect	17
	752.6	Hypospadias & epispadias	15
	745.5	Ostium secundum atrial septal defect	12
	754.3	Congenital dislocation of hip	11
	752.5	Undescended testicle	8
	755.6	Other anomalies of lower limb including pelvic girdle	8
	750.5	Congenital hypertrophic pyloric stenosis	7
	747.0	Patent ductus arteriosus	6
	755.0	Polydactyly	6
	753.2	Obstructive defects of renal pelvis & ureter	5

Section IV
Most Frequently Reported Major Congenital Malformations by County, 1998-2001
Congenital Malformations Registry

County	ICD-9 Code	Description	Number in 1998-2001
Ulster	752.6	Hypospadias & epispadias	35
	754.3	Congenital dislocation of hip	30
	752.5	Undescended testicle	26
	745.4	Ventricular septal defect	24
	750.5	Congenital hypertrophic pyloric stenosis	20
	745.5	Ostium secundum atrial septal defect	17
	753.2	Obstructive defects of renal pelvis & ureter	17
	755.0	Polydactyly	16
	754.5	Varus deformities of feet	11
	758.0	Down syndrome	9
Warren	745.5	Ostium secundum atrial septal defect	11
	752.5	Undescended testicle	8
	745.4	Ventricular septal defect	7
	752.8	Other specified anomalies of genital organs	6
	753.2	Obstructive defects of renal pelvis & ureter	6
	754.3	Congenital dislocation of hip	6
	750.5	Congenital hypertrophic pyloric stenosis	5
	752.6	Hypospadias & epispadias	4
	754.5	Varus deformities of feet	4
	742.3	Congenital hydrocephalus	3
	746.8	Other specified anomalies of heart	3
	748.3	Other anomalies of larynx, trachea, & bronchus	3
	755.0	Polydactyly	3
	758.0	Down syndrome	3
Washington	750.5	Congenital hypertrophic pyloric stenosis	11
	752.5	Undescended testicle	8
	745.4	Ventricular septal defect	6
	752.6	Hypospadias & epispadias	6
	755	Polydactyly	6
	753.2	Obstructive defects of renal pelvis & ureter	5
	745.5	Ostium secundum atrial septal defect	4
	753.1	Cystic kidney disease	3
	754.5	Varus deformities of feet	3
	746.8	Other specified anomalies of heart	2
	747.1	Coarctation of aorta	2
	749.2	Cleft palate with cleft lip	2
	752.8	Other specified anomalies of genital organs	2
	754.7	Other deformities of feet	2
	755.1	Syndactyly	2
	755.2	Reduction deformities of upper limb	2
	756.7	Anomalies of abdominal wall	2

Section IV
Most Frequently Reported Major Congenital Malformations by County, 1998-2001
Congenital Malformations Registry

County	ICD-9 Code	Description	Number in 1998-2001
Wayne	752.6	Hypospadias & epispadias	22
	754.5	Varus deformities of feet	17
	750.5	Congenital hypertrophic pyloric stenosis	13
	746.8	Other specified anomalies of heart	11
	752.5	Undescended testicle	11
	753.2	Obstructive defects of renal pelvis & ureter	10
	745.4	Ventricular septal defect	7
	745.5	Ostium secundum atrial septal defect	7
	745.1	Transposition of great vessels	5
	749.1	Cleft lip	5
	756.0	Anomalies of skull and face bones	5
Westchester			
	752.6	Hypospadias & epispadias	230
	753.2	Obstructive defects of renal pelvis & ureter	200
	752.5	Undescended testicle	155
	745.4	Ventricular septal defect	145
	745.5	Ostium secundum atrial septal defect	125
	755.0	Polydactyly	106
	747.0	Patent ductus arteriosus	83
	754.3	Congenital dislocation of hip	72
	750.5	Congenital hypertrophic pyloric stenosis	71
	758.0	Down syndrome	42
Wyoming	745.5	Ostium secundum atrial septal defect	16
	752.5	Undescended testicle	9
	752.6	Hypospadias & epispadias	9
	745.4	Ventricular septal defect	8
	753.2	Obstructive defects of renal pelvis & ureter	7
	746.8	Other specified anomalies of heart	5
	750.5	Congenital hypertrophic pyloric stenosis	5
	754.3	Congenital dislocation of hip	4
	754.5	Varus deformities of feet	4
	747.1	Coarctation of aorta	3
	749.0	Cleft palate	3
	749.2	Cleft palate with cleft lip	3
Yates	752.6	Hypospadias & epispadias	9
	752.5	Undescended testicle	5
	753.2	Obstructive defects of renal pelvis & ureter	5
	745.5	Ostium secundum atrial septal defect	3
	750.5	Congenital hypertrophic pyloric stenosis	3
	745.4	Ventricular septal defect	2
	754.5	Varus deformities of feet	2

Section IV
Most Frequently Reported Major Congenital Malformations by County, 1998-2001
Congenital Malformations Registry

County	ICD-9 Code	Description	Number in 1998-2001
Yates	754.6	Valgus deformities of feet	2
	277.0	Cystic fibrosis	1
	333.2	Myoclonus	1
	742.3	Congenital hydrocephalus	1
	746.7	Hypoplastic left heart syndrome	1
	746.8	Other specified anomalies of heart	1
	747.3	Anomalies of pulmonary artery	1
	748.3	Other anomalies of larynx, trachea, & bronchus	1
	748.5	Agenesis, hypoplasia & dysplasia, lung	1
	750.3	Tracheoesophageal fistula, esophageal atresia & stenosis	1
	751.1	Atresia & stenosis of small intestine	1
	751.3	Hirschprung's disease & other functional disorders of colon	1
	752.4	Anomalies of cervix, vagina & external female genitalia	1
	754.0	Deformities of skull, face, & jaw	1
	754.3	Congenital dislocation of hip	1
	754.8	Other specified nonteratogenic anomalies	1
	755.0	Polydactyly	1
	755.1	Syndactyly	1
	755.5	Other anomalies of upper limb including shoulder girdle	1
	755.6	Other anomalies of lower limb including pelvic girdle	1
	756.0	Anomalies of skull and face bones	1
	756.6	Anomalies of diaphragm	1
	758.1	Patau syndrome	1
	758.6	Gonadal dysgenesis	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) and the Texas Birth Defects Monitoring Division (TBDMD).[1] 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.) With the exception of Fetal Alcohol Syndrome (FAS) which is followed up to six years in Texas, these programs follow children through one year of age. The CMR follows children through two years; however, more than 95% of cases are reported in the first year. Most of the malformations in this table are recognized at birth. The exceptions are fetal alcohol syndrome and some cardiac malformations.

Several malformations, including tetralogy of fallot, esopheal atresia/tracheoesophageal fistula, pyloric stenosis, and cleft palate without cleft lip have similar prevalence rates among the registries indicating that the CMR's surveillance system is effective in case ascertainment for these defects.

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

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

There are also racial and ethnic differences in the rates of specific birth defects. A report from the California Birth Defects Monitoring Program (CBDMP) for the years 1983-1990 showed that whites had the highest pyloric stenosis rate, hispanics the highest rates of neural tube defects and blacks had a higher rate of heart defects than the other races [2]. There may also be true geographic differences. A comparison of birth defect prevalences between the MACDP and CBDMP for the years 1983-1988 that adjusted for race, sex and maternal age showed regional differences in arm, hand and limb reduction defects [3].

Although there are differences in the rates of some specific defects presented in Table 5, these differences are not substantive.

Section V
Comparison of Selected Malformation Prevalence With Two Other Birth Defects Registries

Malformations	CMR 1997-2001	MACDP[1] 1997-2001	TBDMD[1] 1997-2001
Anencephalus	0.4	3.4	3.0
Spina bifida without anencephalus	2.3	3.6	4.0
Hydrocephalus without spina bifida	7.2	8.2	7.4
Encephalocele	0.7	1.4	1.0
Microcephalus	4.6	7.9	6.5
Anophthalmia/microphthalmia	0.7	2.8	2.8
Common truncus	0.6	0.8	0.8
Transposition of great arteries	3.6	5.4	4.9
Tetralogy of Fallot	4.2	3.7	3.1
Ventricular septal defect	34.6	39.8	42.3
Hypoplastic left heart syndrome	1.8	3.1	2.0
Coarctation of aorta	3.8	5.5	4.4
Choanal atresia	1.5	1.5	1.2
Cleft palate without cleft lip	5.8	7.1	6.1
Cleft lip with and without cleft palate	7.1	8.4	11.1
Esophageal atresia/tracheoesophageal fistula	2.2	2.4	2.1
Rectal/large intestine atresia/stenosis	3.9	4.0	4.8
Pyloric stenosis	17.1	13.4	19.3
Hirschsprung's disease (congenital megacolon)	2.0	2.3	1.2
Biliary atresia	0.9	0.8	0.7
Renal agenesis/hypoplasia	2.9	4.4	5.2
Bladder exstrophy	0.2	0.1	0.2
Hypospadias and epispadias	33.1	32.7	27.8
Reduct deform of upper limb	1.7	4.0	4.2
Reduct deform of lower limb	1.2	1.7	1.9
Diaphragmatic hernia	1.2	2.3	2.5
Omphalocele	1.1	2.5	2.3
Gastroschisis	1.2	2.2	4.0
Down syndrome	10.6	13.0	12.2
Trisomy 13	0.8	1.4	1.2
Trisomy 18	0.2	2.6	2.3
Fetal alcohol syndrome	1.1	1.4	0.2
Amniotic bands	0.2	1.5	0.8

References

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

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. *Morbidity and Mortality Weekly Report* 1993; 42(No. SS-1):1-7.

Section VI
Current Topics

NTD Surveillance and Trends

In 1992, the United States Public Health Service first recommended that all women capable of becoming pregnant take folic acid to prevent neural tube defects (NTD).[1] This recommendation was based on study results including clinical trials showing that folic acid supplementation taken before becoming pregnant and through the first trimester of pregnancy reduced the risk of having an infant affected with an NTD by 50% for both first pregnancies and those following NTD-affected pregnancies [2,3] Recognizing its importance, the fortification of folic acid to grain products as a public health intervention was mandated by the United States Food and Drug Administration in 1998. Studies that involved at least twenty-three population based surveillance systems across the United States, including New York State, looked at rates of NTD-affected pregnancies before and after the mandated fortification and concluded that there was a decline in NTD-affected pregnancies, especially those diagnosed with spina bifida.[4]

New York State has mounted a multi-faceted approach to this public health problem. A major campaign to get information to physicians and the public was initiated in 1996. Knowledge about and use of folic acid is tracked with the PRAMS (Pregnancy Risk Assessment Monitoring System) and the BRFSS (Behavioral Risk Factor Surveillance System), and New York's NTD surveillance system allows us to monitor changes in NTD occurrence.

Since 1996, the Congenital Malformations Registry (CMR) has had an intensive surveillance system for NTDs in seven counties of the Lower Hudson Valley (HSA 6) where about 30,000 births occur yearly. A major problem with accurately describing the prevalence of NTD-affected pregnancies is that many surveillance systems only ascertain live born infants. This presents a problem since many NTD-affected pregnancies are diagnosed prenatally; therefore, pregnancies resulting in fetal death or elective termination are not counted. Widespread use of prenatal ultrasound and blood tests has led to increased detection and termination of NTD-affected pregnancies.[6] The CMR NTD surveillance actively ascertains cases, using hospitals, health care providers and termination facilities. The surveillance goes back retrospectively to 1990, and since 1996 ascertainment has been prospective. For 1990 to 2001, a total of 231 cases of NTD-affected pregnancies have been identified. For the same time period, only 114 cases were reported to the CMR, which includes only live born cases. The percent of NTD-affected pregnancies that have been terminated in HSA 6 is shown in Figure 1. Termination of NTD-affected pregnancies continued to increase from 23% in 1990 to over 50% in 2003 in that region of the state. As terminations increase, ascertainment by passive surveillance systems like the CMR will greatly underestimate the occurrence of neural tube defects.

The rates per 1,000 births of anencephaly, spina bifida, and total NTD found by the active surveillance in HSA Region 6 and based on CMR reports are displayed in Figures 2, 3 and 4. The difference in ascertainment of these defects by the two surveillance systems is quite evident. Also presented are three-year moving average trend lines. These indicate that the rates of NTD-affected pregnancies have decreased by 22% over time. Since New York's campaign to educate the medical community and women of childbearing age of the benefits of folic acid consumption coincided with mandated fortification of grain products, it is difficult to attribute the decrease seen in NTD-affected pregnancies to one prevention measure. However, since NTDs are potentially preventable and NTD-affected pregnancies are still occurring, continued education and surveillance will only benefit the publc.

Figure 1
Percent of Affected Pregnancies Terminated, Lower Hudson Valley
Neural Tube Defect Surveillance System, 1990 to 2003

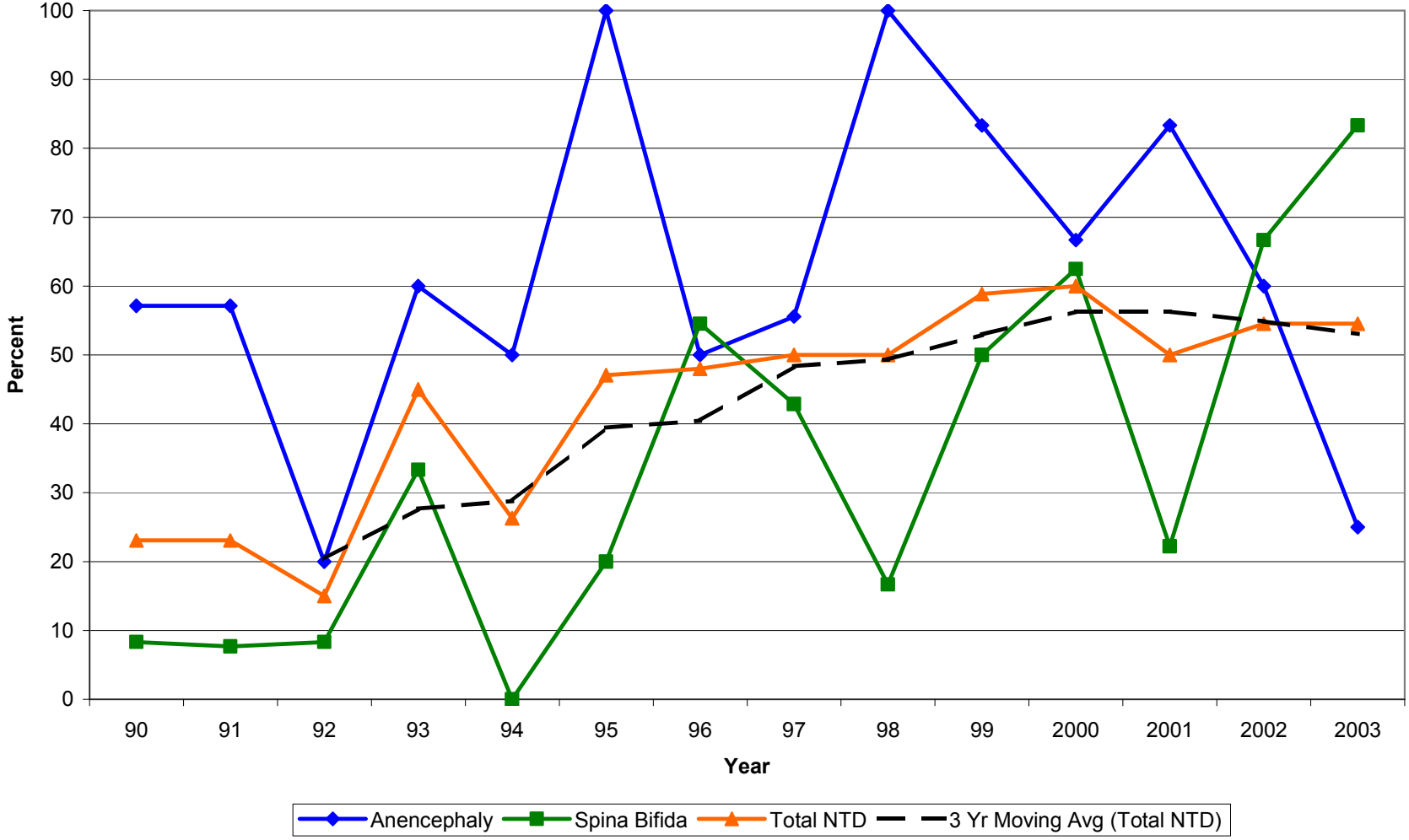


Figure 2
Anencephaly, Lower Hudson Valley
Neural Tube Defect Surveillance System, 1990 to 2003
Congenital Malformations Registry, 1990 to 2001

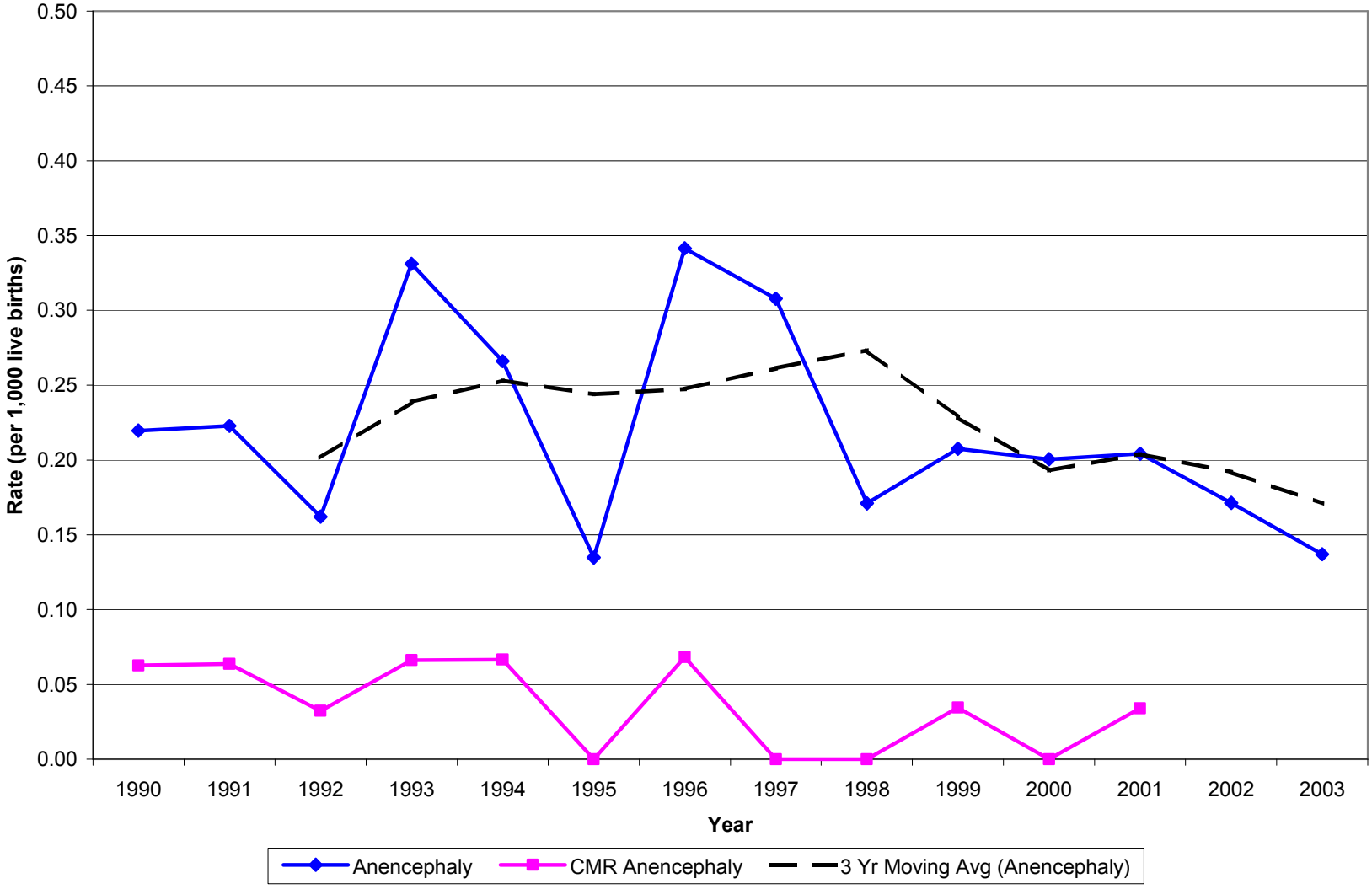
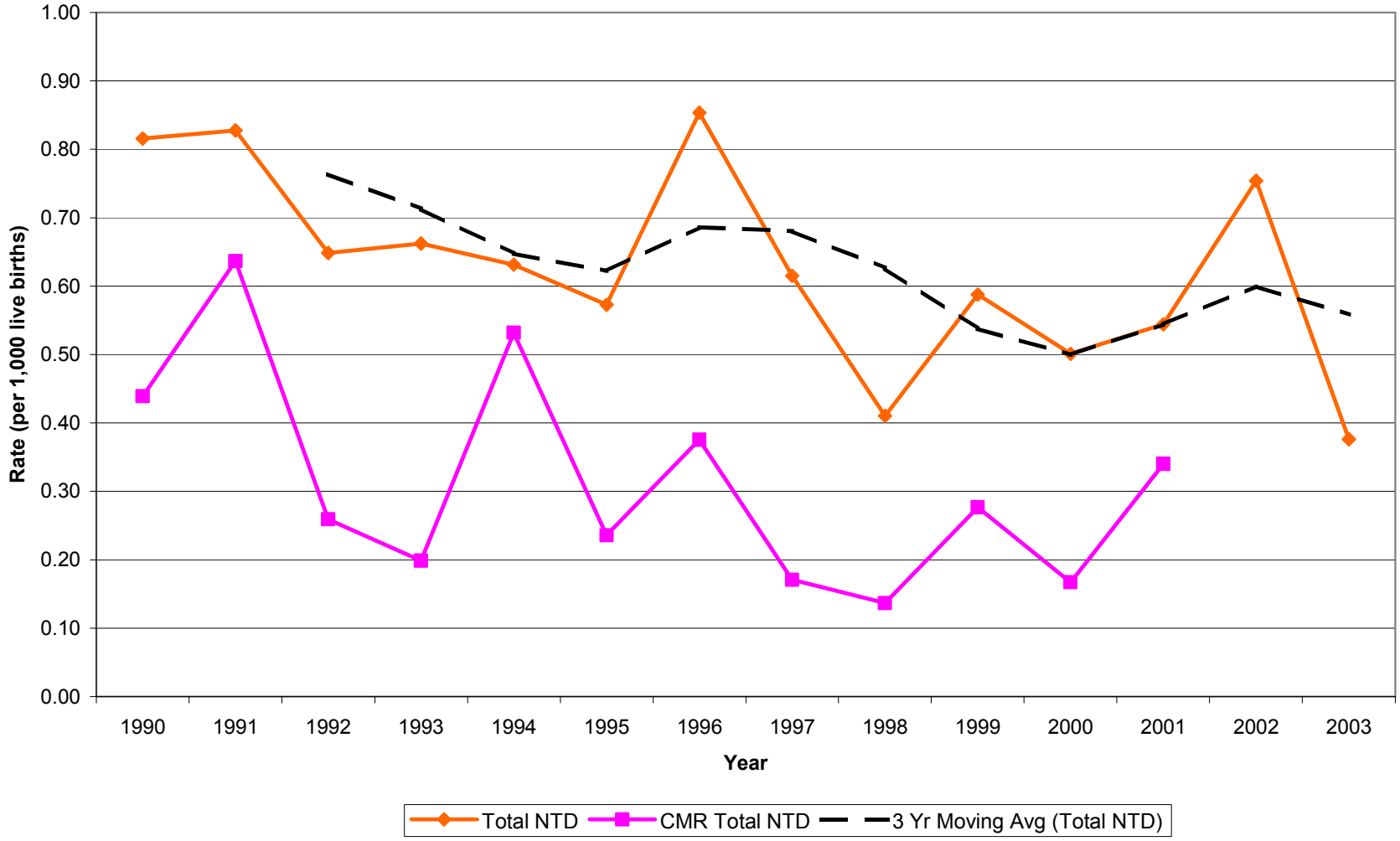


Figure 3
Spina Bifida, Lower Hudson Valley
Neural Tube Defect Surveillance System, 1990 to 2003
Congenital Malformations Registry, 1990 to 2001

Figure 4
Total Neural Tube Defects, Lower Hudson Valley
Neural Tube Defect Surveillance System, 1990 to 2003
Congenital Malformations Registry, 1990 to 2001



References

[1]Center for Disease Control. Recommendations for the use of folic acid to reduce the number of cases of spina bifida and other neural tube defects. MMWR 1992;41:1-7.

[2]MRC Vitamin Study Research Group. Prevention of neural tube defects: results of the Medical Research Council vitamin study. Lancet 1991;338(8760):131-37.

[3]Czeizel AE, Dudas I. Prevention of the first occurrence of neural-tube defects by periconceptional vitamin supplementation. NEJM 1992;327(26):1832-35.

[4]Williams LJ, Mai CT, Edmonds LD, Shaw GM, Kirby RS, Hobbs CA, Sever LE, Miller LA, Meaney FJ, Levitt M. Prevalence of spina bifida and anencephaly during the transition to mandatory folic acid fortification in the United States. Teratology 2002;66:33-9.

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[6]Peller AJ, Westgate MN, Holmes LB. Trends in congenital malformations, 1974-1999: effect of prenatal diagnosis and elective termination. Obstetrics & Gynecology 2004;104(5):957-64.

APPENDICES

Appendix 1

Report Form, Congenital Malformations Registry

Congenital Malformations Registry
Confidential Case Report

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

Type or print clearly using blue or black ink.

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

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

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

Address Street City State Zip Code

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

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

Hispanic ☐ Yes ☐ No

Plurality
☐ Single ☐ Twin ☐ Triplet Other, specify _____

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

Born at this facility
☐ Yes ☐ No

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

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

Diagnostic Information

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

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

Parents' Information

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

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

Father's Name Last First M.I.

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

Type or print clearly using blue or black ink.

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

Mail completed form in sealed envelope to:

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

Appendix 2

Classification of Codes

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

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

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

Major Malformations

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

*See list of minor and excluded codes

Minor Malformations

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

Exclusions

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

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Appendix 3

Birth Certificate Matching

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

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

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

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

Appendix 4

BPA Codes

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

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

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

Appendix 5

Glossary of Terms*

Agenesis Absence of part(s) of the body.

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

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

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

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

Anotia A congenital absence of one or both ears.

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

Atresia Imperforation; absence or closure of a normal opening.

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

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

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

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

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

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

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

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

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

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

Congenital Existing at or dating from birth.

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

Limb defects See Reduction deformities.

Meninges Membranes that cover the brain and spinal cord.

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

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

Trisomy 13 See Patau Syndrome.

Trisomy 18 See Edwards Syndrome.

Trisomy 21 See Down Syndrome.

Truncus Arteriosus See Common Truncus.

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

*Courtesy of the Texas Birth Defects Monitoring Division February 1999