

10. BIRTH DEFECTS

Birth defects among stillborn and liveborn infants

A birth defect is any structural defect detected during pregnancy or at birth, excluding birth injuries and minor anomalies such as skin tags, positional talipes, birthmarks, or clicky hips. Descriptions of some common birth defects are shown in Appendix 1. A list of common exclusions is shown in Appendix 2.

From 1 January 1998, doctors, hospitals and laboratories are required to notify birth defects detected during pregnancy, at birth, or up to one year of life under the *NSW Public Health Act 1991*. Information reported is included in the NSW Birth Defects Register (BDR). The quality of information received by the BDR has improved since 1998, particularly in relation to pregnancy outcome.

This chapter reports birth defects detected during pregnancy or in the first year of life for 1999–2004 and birth defects detected during pregnancy or at birth for 2005.

Trends in reported birth defects

Between 1999 and 2004, the reported number of infants with birth defects has remained stable at just over 2 per cent (Table 112). In 2005, 817 cases of birth defects detected during pregnancy or at birth were reported.

Birth defects by diagnostic category

The most common categories of birth defects for births of more than 20 weeks gestation or with a birth weight greater

TABLE 112

BIRTH DEFECT CASES, NSW 1999–2005#

Year	Birth defect cases	Births	Rate/1,000 births
1999	1828	86468	21.1
2000	1858	87140	21.3
2001	1775	85286	20.8
2002	1739	85398	20.4
2003	1761	85853	20.5
2004	1758	85016	20.7
2005	817	89840	9.1

Source: NSW Birth Defects Register. Centre for Epidemiology and Research, NSW Department of Health.

For 1999–2004, cases reported during pregnancy and up to one year of age are included. For 2005, cases reported during pregnancy or at birth are reported.

than 400 grams are presented in Table 113. Birth defects are classified using the British Paediatric Association (BPA) Classification of Diseases, which is primarily organised by body system. For infants with more than one defect, each defect is counted separately. The number of birth defects reported therefore exceeds the number of affected infants.

In 1999–2005, defects of the cardiovascular system were most commonly reported, followed by defects of the musculoskeletal system and defects of the genito-urinary system (Table 113). This is a similar pattern to previous years. In 2004, the overall rate of defects was similar to the previous 5 years (37.3 versus 36.7 per 1,000).

TABLE 113

BIRTH DEFECTS AMONG STILLBIRTHS AND LIVE BIRTHS BY DIAGNOSTIC CATEGORY, NSW 1999–2005

Diagnostic category	No. defects			1999–2005	Rate/1,000 births			
	1999–2003	2004	2005		1999–2003	2004	2005	1999–2005
Defects of nervous system								
Anencephaly	49	7	10	66	0.1	0.1	0.1	0.1
Spina Bifida	137	23	20	180	0.3	0.3	0.2	0.3
Encephalocele	28	8	3	39	0.1	0.1	0.0	0.1
Microcephaly	111	24	5	140	0.3	0.3	0.1	0.2
Congenital hydrocephalus	174	43	22	239	0.4	0.5	0.2	0.4
Other nervous system defects	334	61	19	414	0.8	0.7	0.2	0.7
TOTAL	833	166	79	1078	1.9	2.0	0.9	1.8
Defects of eye								
Anophthalmos–microphthalmos	54	20	4	78	0.1	0.2	0.0	0.1
Buphthalmos–congenital glaucoma	27	4	3	34	0.1	0.0	0.0	0.1
Congenital cataract	75	15	5	95	0.2	0.2	0.1	0.2
Other eye defects	161	40	12	213	0.4	0.5	0.1	0.4
TOTAL	317	79	24	420	0.7	0.9	0.3	0.7
Defects of ear, face and neck								
Absence/ stricture auditory canal	54	7	10	71	0.1	0.1	0.1	0.1
Absent auricle	8	1	1	10	0.0	0.0	0.0	0.0
Defects of face and neck	40	8	3	51	0.1	0.1	0.0	0.1
Other ear defects	79	11	8	98	0.2	0.1	0.1	0.2
TOTAL	181	27	22	230	0.4	0.3	0.2	0.4
Defects of cardiovascular system								
Transposition of great vessels	215	44	39	298	0.5	0.5	0.4	0.5
Tetralogy of Fallot	147	31	13	191	0.3	0.4	0.1	0.3
Ventricular septal defect	853	178	79	1110	2.0	2.1	0.9	1.8
Atrial septal defect	821	172	67	1060	1.9	2.0	0.7	1.8

TABLE 113 (continued)

BIRTH DEFECTS AMONG STILLBIRTHS AND LIVE BIRTHS BY DIAGNOSTIC CATEGORY, NSW 1999–2005#

Diagnostic category	No. defects				Rate/1,000 births			
	1999–2003	2004	2005	1999–2005	1999–2003	2004	2005	1999–2005
Defects of cardiovascular system (cont.)								
Heart valve defects	595	108	52	755	1.4	1.3	0.6	1.2
Patent ductus arteriosus > 37 weeks	446	106	50	602	1.0	1.2	0.6	1.0
Coarctation of aorta	199	43	20	262	0.5	0.5	0.2	0.4
Other defects of aorta	107	22	14	143	0.2	0.3	0.2	0.2
Defects of pulmonary artery	140	30	21	191	0.3	0.4	0.2	0.3
Other cardiovascular defects	754	154	97	1005	1.8	1.8	1.1	1.7
TOTAL	4277	888	452	5617	9.9	10.4	5.0	9.3
Defects of respiratory system								
Defects of nose	66	9	6	81	0.2	0.1	0.1	0.1
Defects of larynx, trachea and bronchus	45	12	1	58	0.1	0.1	0.0	0.1
Defects of lung	79	18	8	105	0.2	0.2	0.1	0.2
TOTAL	190	39	15	244	0.4	0.5	0.2	0.4
Defects of gastrointestinal system								
Cleft palate only	391	84	67	542	0.9	1.0	0.7	0.9
Cleft lip only	147	34	19	200	0.3	0.4	0.2	0.3
Cleft palate and cleft lip	228	42	31	301	0.5	0.5	0.3	0.5
Oesophageal atresia only	11	2	3	16	0.0	0.0	0.0	0.0
Oesophageal atresia with TOF	82	12	11	105	0.2	0.1	0.1	0.2
Tracheo-oesophageal fistula (TOF) only	22	2	2	26	0.1	0.0	0.0	0.0
Atresia-stenosis of small intestine	144	18	15	177	0.3	0.2	0.2	0.3
Atresia-stenosis of anus	127	24	20	171	0.3	0.3	0.2	0.3
Other gastrointestinal defects	468	96	20	584	1.1	1.1	0.2	1.0
TOTAL	1620	314	188	2122	3.8	3.7	2.1	3.5
Defects of genitourinary system								
Defects of female genitals	43	17	5	65	0.1	0.2	0.1	0.1
Undescended testis	313	63	9	385	0.7	0.7	0.1	0.6
Hypospadias	865	150	111	1126	2.0	1.8	1.2	1.9
Epispadias	13	0	0	13	0.0	0.0	0.0	0.0
Chordee	136	22	19	177	0.3	0.3	0.2	0.3
Indeterminate sex-ambiguous genitalia	51	11	4	66	0.1	0.1	0.0	0.1
Renal agenesis-dysgenesis	193	42	21	256	0.4	0.5	0.2	0.4
Obstructive defects of renal pelvis and ureter	716	152	29	897	1.7	1.8	0.3	1.5
Other genitourinary system defects	599	126	57	782	1.4	1.5	0.6	1.3
TOTAL	2929	583	255	3767	6.8	6.9	2.8	6.2
Defects of musculoskeletal system								
Congenital dislocation of the hips	670	133	34	837	1.6	1.6	0.4	1.4
Talipes equinovarus	326	73	21	420	0.8	0.9	0.2	0.7
Polydactyly	482	96	78	656	1.1	1.1	0.9	1.1
Syndactyly	90	19	21	130	0.2	0.2	0.2	0.2
Reduction deformities of limbs	234	51	45	330	0.5	0.6	0.5	0.5
Craniosynostosis	304	41	3	348	0.7	0.5	0.0	0.6
Diaphragmatic hernia	127	31	21	179	0.3	0.4	0.2	0.3
Exomphalos	72	16	11	99	0.2	0.2	0.1	0.2
Gastroschisis	93	16	17	126	0.2	0.2	0.2	0.2
Other musculoskeletal defects	818	159	95	1072	1.9	1.9	1.1	1.8
TOTAL	3216	635	346	4197	7.5	7.5	3.9	6.9
Defects of the integumentary system								
Cystic hygroma	49	9	7	65	0.1	0.1	0.1	0.1
Chromosomal defects								
Trisomy 21	505	98	49	652	1.2	1.2	0.5	1.1
Trisomy 13	36	7	6	49	0.1	0.1	0.1	0.1
Trisomy 18	89	18	12	119	0.2	0.2	0.1	0.2
Turner syndrome	69	13	8	90	0.2	0.2	0.1	0.1
Other chromosomal defects	293	67	31	391	0.7	0.8	0.3	0.6
TOTAL	992	203	106	1301	2.3	2.4	1.2	2.2
Situs inversus								
Situs inversus	27	4	3	34	0.1	0.0	0.0	0.1
Congenital malformation syndromes								
Congenital malformation syndromes	190	44	19	253	0.4	0.5	0.2	0.4
Congenital cytomegalovirus infection								
Congenital cytomegalovirus infection	2	1	0	3	0.0	0.0	0.0	0.0
Congenital toxoplasmosis								
Congenital toxoplasmosis	1	0	0	1	0.0	0.0	0.0	0.0
Non-immune hydrops foetalis								
Non-immune hydrops foetalis	115	23	14	152	0.3	0.3	0.2	0.3
Other and unspecified birth defects								
Other and unspecified birth defects	508	98	15	621	1.2	1.2	0.2	1.0
TOTAL	15770	3170	1557	20497	36.7	37.3	17.3	33.9

Source: NSW Birth Defects Register. Centre for Epidemiology and Research, NSW Department of Health.

For 1999–2004, cases reported during pregnancy and up to one year of age are included. For 2005, cases reported during pregnancy or at birth are reported.

Infant characteristics

In the period 1999–2005, a single defect was reported in 64.1 per cent of infants, 2 defects in 18.0 per cent, 3 defects in 8.2 per cent, and 4 or more defects in 9.8 per cent of cases.

The sex was male in 57.8 per cent of infants, female in 41.8 per cent, indeterminate in 0.3 per cent of infants, and was not stated for 0.2 per cent.

Birth defects were more common in preterm and post-term infants than infants born at term (Table 114). Birth defects were also more common in infants born of a multiple

pregnancy than a singleton pregnancy: in 1999–2005, 1.9 per cent of singleton babies, 2.5 per cent of twins, and 4.0 per cent of triplets were born with a birth defect.

About 11 per cent of infants born with birth defects died in the perinatal period, over half of which were stillbirths (Table 115). These figures comprise all birth defect cases, including those where the cause of death may not be directly related to the birth defect(s). By comparison, the perinatal mortality rate among all births reported to the NSW Midwives Data Collection was less than one per cent in 2005 (see Chapter 4).

TABLE 114

BIRTH DEFECT CASES BY GESTATIONAL AGE, NSW 1999–2005[#]

Gestational age (weeks)	1999–2003		2004		Year 2005		1999–2005		Rate/1,000 births
	No.	%	No.	%	No.	%	No.	%	
20–27	569	6.3	125	7.1	79	9.7	773	6.7	184.5
28–31	262	2.9	65	3.7	19	2.3	346	3.0	76.9
32–36	1065	11.9	233	13.3	126	15.4	1424	12.3	41.1
37–41	6498	72.5	1241	70.6	580	71.0	8319	72.1	15.2
42+	194	2.2	31	1.8	13	1.6	238	2.1	17.4
Not stated	373	4.2	63	3.6	0	0.0	436	3.8	–
TOTAL	8961	100.0	1758	100.0	817	100.0	11536	100.0	19.1

Source: NSW Birth Defects Register. Centre for Epidemiology and Research, NSW Department of Health.

[#] For 1999–2004, cases reported during pregnancy and up to one year of age are included. For 2005, cases reported during pregnancy or at birth are reported.

TABLE 115

BIRTH DEFECT CASES BY PREGNANCY OUTCOME, NSW 1999–2005[#]

Pregnancy outcome	1999–2003		2004		Year 2005		1999–2005	
	No.	%	No.	%	No.	%	No.	%
Stillbirth	547	6.1	125	7.1	91	11.1	763	6.6
Liveborn/neonatal death	384	4.3	68	3.9	45	5.5	497	4.3
Liveborn/ postneonatal death	76	0.8	9	0.5	3	0.4	88	0.8
Liveborn surviving	7954	88.8	1556	88.5	678	83.0	10188	88.3
TOTAL	8961	100.0	1758	100.0	817	100.0	11536	100.0

Source: NSW Birth Defects Register. Centre for Epidemiology and Research, NSW Department of Health.

[#] For 1999–2004, cases reported during pregnancy and up to one year of age are included. For 2005, cases reported during pregnancy or at birth are reported. Postneonatal deaths are likely to be under-reported.

Maternal characteristics

After 35 years of age, the incidence of birth defects increased with increasing maternal age (Table 116). While the rate of birth defects is higher in older women, the majority of births occur in younger women. In 1999–2005, 73.5 per cent of babies with birth defects were born to women aged less than 35 years.

In 1999–2005, 292 babies of Aboriginal or Torres Strait Islander mothers were reported to have birth defects. The rate of birth defects among these babies was 18.9 per 1,000 compared with 18.0 per 1,000 for non-Aboriginal mothers.

TABLE 116

BIRTH DEFECT CASES BY MATERNAL AGE, NSW 1999–2005#

Maternal age (years)	1999–2005		2004		Year 2005		1999–2005		Rate/1,000 births
	No.	%	No.	%	No.	%	No.	%	
Under 20	380	4.2	68	3.9	40	4.9	488	4.2	19.1
20–24	1270	14.2	238	13.5	135	16.5	1643	14.2	18.2
25–29	2454	27.4	436	24.8	192	23.5	3082	26.7	17.4
30–34	2485	27.7	538	30.6	241	29.5	3264	28.3	16.6
35–39	1323	14.8	282	16.0	152	18.6	1757	15.2	18.5
40–44	330	3.7	62	3.5	52	6.4	444	3.8	24.0
45+	21	0.2	7	0.4	5	0.6	33	0.3	37.5
Not stated	698	7.8	127	7.2	0	0.0	825	7.2	–
TOTAL	8961	100.0	1758	100.0	817	100.0	11536	100.0	19.1

Source: NSW Birth Defects Register. Centre for Epidemiology and Research, NSW Department of Health.

For 1999–2004, cases reported during pregnancy and up to one year of age are included. For 2005, cases reported during pregnancy or at birth are reported.

Birth defects among terminations of pregnancy, spontaneous abortions and unknown outcomes of pregnancy

In the period 1999–2005, on average about 300 terminations of pregnancy per year were reported to the NSW Birth Defects Register (Table 117). To date, 152 terminations of pregnancy have been reported to the Register for 2005. This number is expected to increase as outcomes for mothers with defects detected during pregnancy in 2005 continue to be reported. Of the 1,881 terminations of pregnancy reported in 1999–2005, 1,435 (76.3 per cent)

were associated with a chromosomal abnormality, the most common of which was Trisomy 21 (Down syndrome), and 212 (11.3 per cent) were associated with a neural tube defect (Tables 117 and 118).

For spontaneous abortions, cytogenetic analysis is only carried out in cases of habitual abortion; the numbers presented, therefore, underestimate the number of spontaneous abortions that occur due to birth defects. Descriptions of some diagnostic terms used here are included in Appendix 1.

TABLE 117

PREGNANCIES WITH FETUSES AFFECTED BY BIRTH DEFECTS AND RESULTING IN SPONTANEOUS ABORTION, TERMINATION OF PREGNANCY OR UNKNOWN OUTCOME, NSW 1999–2005

Pregnancy outcome	1999	2000	2001	2002	Year 2003	2004	2005	1999–2005
	No.	No.	No.	No.	No.	No.	No.	
Spontaneous abortion	119	124	171	202	232	298	308	1454
Termination of pregnancy								
less than 20 weeks gestation	310	262	257	285	312	303	152	1881
Unknown outcome	16	22	19	7	18	41	0	123
TOTAL	445	408	447	494	562	642	460	3458

Source: NSW Birth Defects Register. Centre for Epidemiology and Research, NSW Department of Health.

TABLE 118

BIRTH DEFECTS AMONG SPONTANEOUS ABORTIONS, TERMINATIONS OF PREGNANCY AND UNKNOWN OUTCOME OF PREGNANCY BY DIAGNOSTIC CATEGORY, NSW 1999–2005

Diagnostic category	1999–2003			Year 2004			2005			1999–2005		
	Spont. abortion No.	Termination of pregnancy less than 20 weeks gestation No.	Unknown outcome No.	Spont. abortion No.	Termination of pregnancy less than 20 weeks gestation No.	Unknown outcome No.	Spont. abortion No.	Termination of pregnancy less than 20 weeks gestation No.	Unknown outcome No.	Spont. abortion No.	Termination of pregnancy less than 20 weeks gestation No.	Unknown outcome No.
Defects of nervous system												
Neural tube defects	5	169	3	1	21	1	1	22	7	212	4	
Other nervous system defects	5	122	6	1	22	0	2	23	8	167	6	
TOTAL	10	291	9	2	43	1	3	45	15	379	10	
Defects of eye	0	3	0	0	0	0	0	2	0	5	0	
Defects of ear, face and neck	1	10	1	0	2	0	0	3	1	15	1	
Defects of cardiovascular system	11	180	7	1	28	2	4	70	16	278	9	
Defects of respiratory system	2	20	3	0	4	1	2	9	4	33	4	
Defects of gastrointestinal system	7	121	4	2	25	0	2	29	11	175	4	
Defects of musculoskeletal system	25	289	9	7	61	2	3	58	35	408	11	
Defects of genitourinary system	11	148	4	2	27	0	2	17	15	192	4	
Defects of integumentary system	0	2	0	1	0	0	0	0	1	2	0	
Cystic hygroma	9	99	3	7	30	2	4	8	20	137	5	
Chromosomal defects												
Trisomy 21	64	538	22	26	132	15	26	49	116	719	37	
Trisomy 13	35	79	11	9	16	3	18	8	62	103	14	
Trisomy 18	43	193	7	16	37	2	11	11	70	241	9	
Turner syndrome	81	91	2	35	23	7	39	9	155	123	9	
Other chromosomal defects	609	200	26	206	36	10	212	13	1027	249	36	
TOTAL	832	1101	68	292	244	37	306	90	1430	1435	105	
Situs inversus	0	4	0	0	0	0	0	1	0	5	0	
Congenital malformation syndromes	2	11	0	0	0	0	0	3	2	14	0	
Non-immune hydrops foetalis	7	48	3	3	11	0	3	7	13	66	3	
Other and unspecified birth defects	4	28	3	1	6	1	0	9	5	43	4	
TOTAL	921	2355	114	318	481	46	329	351	1568	3187	160	

Source: NSW Birth Defects Register. Centre for Epidemiology and Research, NSW Department of Health.

Trends in selected birth defects

Trends in a selection of common birth defects are shown in Table 119 and Figures 14 to 17. For 1999–2004, malformations reported up to one year of age are included; for 2005, malformations reported during pregnancy or at birth are included.

The reported number of liveborn and stillborn infants with neural tube defects was 85 in 1999 and 58 in 2004, and 54 have been reported for 2005 to date. The number of reported terminations of pregnancy was 44 in 1999, 21 in 2004, and 21 in 2005 (Figure 14).

Over the period 1999–2005, the number of cases of isolated cleft palate ranged from 60 to 88 per year, and for total cleft lip (including cases of cleft lip and cleft palate) from 71 to 88 per year. Termination of pregnancy was usually associated with other defects such as neural tube defects, chromosomal abnormalities, or multiple abnormalities in addition to the cleft lip and/or cleft palate.

The reported number of liveborn and stillborn infants with chromosomal defects was 191 in 1999 and 202 in 2004, and the number of reported terminations of pregnancy associated with chromosomal defects rose from 221 in 1999 to 243 in 2004. The number of infants born with Down syndrome was 93 in 1999 and 98 in 2004, while the number of reported terminations of pregnancy associated with Down syndrome rose from 106 in 1999 to 132 in 2004.

In 1999, 37 liveborn infants and 1 stillborn infant had a diaphragmatic hernia, and there were 3 terminations of pregnancy for this condition. In 2004, there were 28 liveborn infants and 3 stillborn infants who had a diaphragmatic hernia, and there were no terminations of pregnancy (Figure 17).

TABLE 119

SELECTED BIRTH DEFECT CASES BY YEAR, NSW 1999–2005[#]

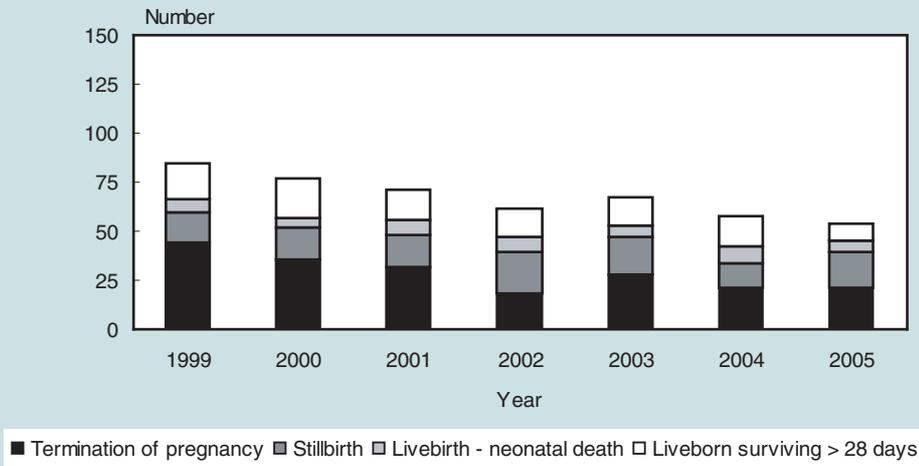
Birth defect	Year													
	1999		2000		2001		2002		2003		2004		2005	
	No.	Rate/ 1,000												
Neural tube defects	85	1.0	77	0.9	71	0.8	62	0.7	67	0.8	58	0.7	54	0.6
Anencephalus	24	0.3	29	0.3	25	0.3	20	0.2	18	0.2	16	0.2	18	0.2
Spina bifida	57	0.7	42	0.5	39	0.5	33	0.4	46	0.5	35	0.4	32	0.4
Encephalocele	8	0.1	13	0.1	8	0.1	10	0.1	7	0.1	8	0.1	5	0.1
Cleft palate	67	0.8	79	0.9	67	0.8	60	0.7	88	1.0	74	0.9	67	0.7
Total cleft lip	84	1.0	71	0.8	88	1.0	76	0.9	85	1.0	80	0.9	57	0.6
Hypospadias	199	2.3	191	2.2	173	2.0	133	1.6	171	2.0	150	1.8	111	1.2
Limb reduction defects	56	0.6	61	0.7	42	0.5	21	0.2	31	0.4	36	0.4	33	0.4
Chromosomal abnormalities	412	4.8	412	4.7	370	4.3	441	5.2	451	5.3	445	5.2	196	2.2
Down syndrome	199	2.3	214	2.5	180	2.1	221	2.6	227	2.6	230	2.7	98	1.1
Renal agenesis and dysgenesis	80	0.9	82	0.9	75	0.9	63	0.7	68	0.8	82	1.0	49	0.5
Exomphalos	17	0.2	28	0.3	22	0.3	22	0.3	20	0.2	28	0.3	15	0.2
Gastroschisis	18	0.2	20	0.2	23	0.3	18	0.2	19	0.2	17	0.2	19	0.2
Diaphragmatic hernia	41	0.5	22	0.3	28	0.3	23	0.3	21	0.2	31	0.4	21	0.2

Source: NSW Birth Defects Register. Centre for Epidemiology and Research, NSW Department of Health.

[#] Includes terminations of pregnancy, stillbirths and livebirths. From 1 January 1998 birth defects became notifiable under the *NSW Public Health Act 1991*. This resulted in increased reporting of birth defects, particularly those associated with termination of pregnancy. For 1999–2004, cases reported during pregnancy and up to one year of age are included. For 2005, cases reported during pregnancy or at birth are reported.

FIGURE 14

NEURAL TUBE DEFECTS: CASES BY YEAR AND PREGNANCY OUTCOME, NSW 1999–2005*

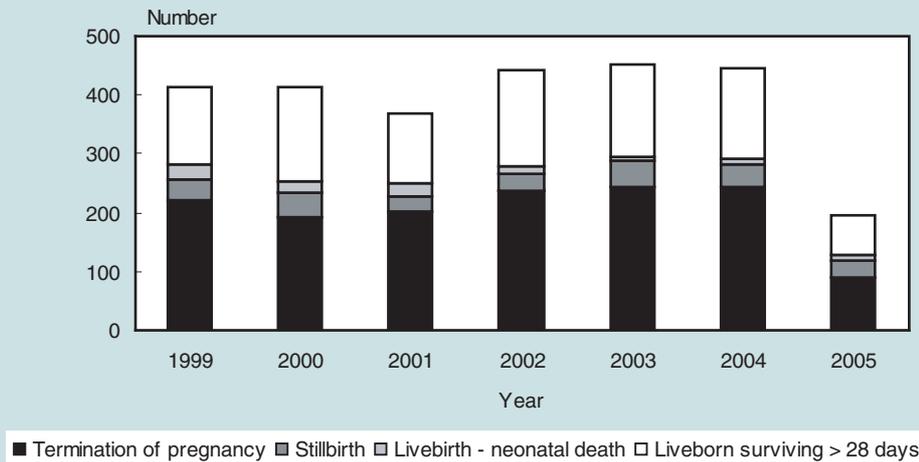


Source: NSW Birth Defects Register. Centre for Epidemiology and Research, NSW Department of Health.

For 1999–2004, cases reported during pregnancy and up to one year of age are included. For 2005, cases reported during pregnancy or at birth are reported.

FIGURE 15

CHROMOSOMAL ABNORMALITIES: CASES BY YEAR AND PREGNANCY OUTCOME, NSW 1999–2005*

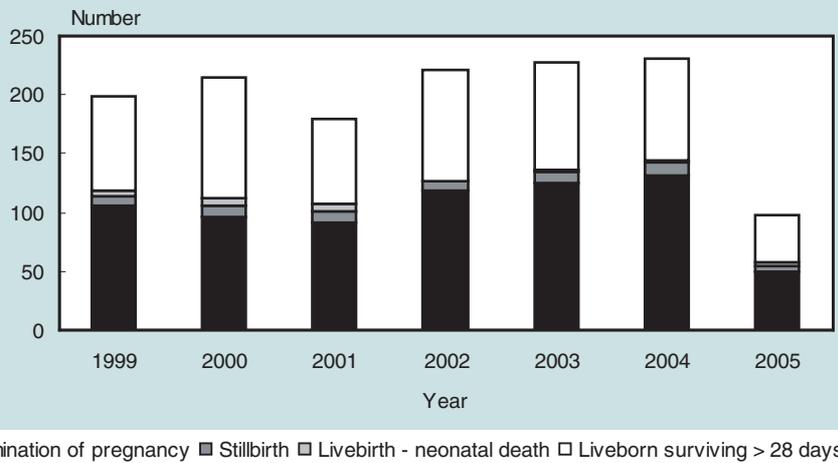


Source: NSW Birth Defects Register. Centre for Epidemiology and Research, NSW Department of Health.

For 1999–2004, cases reported during pregnancy and up to one year of age are included. For 2005, cases reported during pregnancy or at birth are reported.

FIGURE 16

DOWN SYNDROME: CASES BY YEAR AND PREGNANCY OUTCOME, NSW 1999–2005*

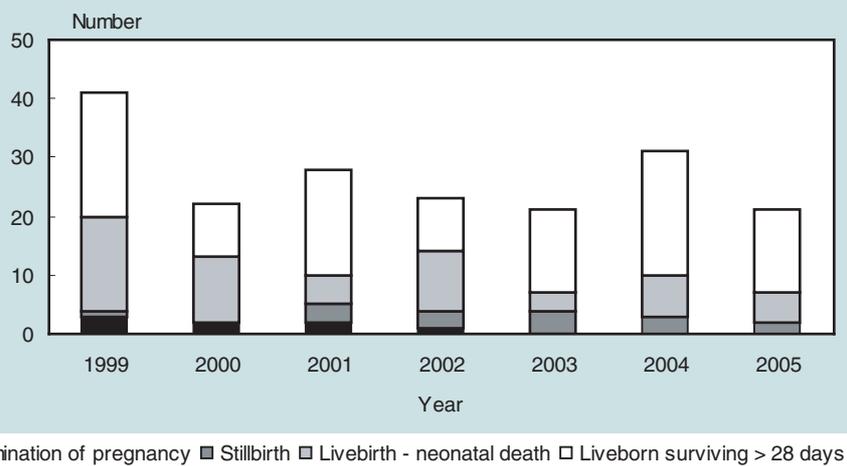


Source: NSW Birth Defects Register. Centre for Epidemiology and Research, NSW Department of Health.

For 1999–2004, cases reported during pregnancy and up to one year of age are included. For 2005, cases reported during pregnancy or at birth are reported.

FIGURE 17

DIAPHRAGMATIC HERNIA: CASES BY YEAR AND PREGNANCY OUTCOME, NSW 1999–2005*



Source: NSW Birth Defects Register. Centre for Epidemiology and Research, NSW Department of Health.

For 1999–2004, cases reported during pregnancy and up to one year of age are included. For 2005, cases reported during pregnancy or at birth are reported.

Birth defects by NSW health areas

Crude rates of reported birth defects for NSW health areas and rates standardised for maternal age are shown in Table 120. The denominator population includes livebirths and stillbirths among NSW residents as reported to the MDC. The rate of birth defects increases with increasing maternal age (Table 116). In order to allow direct comparison of geographic areas, rates have been standardised to the maternal age distribution of births in NSW in 1991.

In interpreting birth defect rates among NSW areas, it should also be noted that infants with birth defects who are born to mothers resident in areas close to interstate borders may be transferred interstate for care and therefore may not be reported to the BDR.

Over the period 1999–2005, standardised rates of reported birth defects were lowest in the Greater Southern Area and highest in the Hunter & New England Area. Review of cases showed slightly increased reported rates of a range of birth defects in the Hunter & New England Area compared to NSW overall including: unstable hips (but not dislocated hips), first degree hypospadias, neural tube defects and a variety of chromosomal abnormalities. The range and pattern of these defects suggests that enumeration of congenital abnormalities is better in the Hunter & New England Health Area compared with NSW as a whole.

TABLE 120

BIRTH DEFECTS IN NSW HEALTH AREAS, 1999–2005#

Health Area	1999–2003			Year			2005			1999–2005			99% confidence intervals
	No.	Crude rate per 1,000 births	Standardised rate per 1,000 births	No.	Crude rate per 1,000 births	Standardised rate per 1,000 births	No.	Crude rate per 1,000 births	Standardised rate per 1,000 births	No.	Crude rate per 1,000 births	Standardised rate per 1,000 births	
Sydney South West	2196	22.6	20.5	436	23.0	20.7	177	8.8	8.4	2809	20.6	18.8	17.8–19.8
South Eastern Sydney & Illawarra	1685	23.9	20.9	295	20.5	17.1	167	11.2	11.7	2147	21.6	19.0	17.8–20.3
Sydney West	1855	23.0	21.2	356	22.1	20.9	154	8.9	8.3	2365	20.7	19.2	18.1–20.3
Northern Sydney & Central Coast	1488	22.5	18.8	303	22.8	19.8	152	10.9	8.7	1943	20.8	17.5	16.1–18.8
Hunter & New England	1375	27.2	25.3	302	30.8	27.5	150	14.2	12.6	1827	25.8	23.7	22.2–25.3
North Coast	482	20.1	19.4	87	18.3	16.6	54	10.7	10.1	623	18.4	17.6	15.8–19.6
Greater Southern	370	17.6	16.1	77	19.8	17.0	38	9.4	9.6	485	16.8	15.3	13.5–17.3
Greater Western	414	20.2	19.2	93	24.2	22.3	43	10.6	10.5	550	19.4	18.4	16.4–20.7
TOTAL NSW	9865	22.9	20.8	1949	22.9	20.6	935	10.4	9.6	12749	21.1	19.1	18.6–19.6

Source: NSW Birth Defects Register. Centre for Epidemiology and Research, NSW Department of Health.

Cases exclude terminations of pregnancy, stillbirths and livebirths where the place of residence is unknown. For 1999–2004, cases reported during pregnancy and up to one year of age are included. For 2005, cases reported during pregnancy or at birth are reported.