

11. BIRTH DEFECTS

Birth defects among stillborn and live born 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). This chapter reports birth defects detected during pregnancy or in the first year of life for 2000–2005 and birth defects detected during pregnancy or at birth for 2006.

Trends in reported birth defects

Between 2000 and 2004, the reported proportion of infants with birth defects has remained stable at about 2 per cent, with a slight decline in 2005 to 1.8 per cent (Table 145). In 2006, 792 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 than 400 grams are presented in Table 146. 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 2000–2006, defects of the cardiovascular system were most commonly reported, followed by defects of the musculoskeletal system and defects of the genitourinary system (Table 146). This is a similar pattern to previous years. In 2005, the overall rate of defects was slightly lower than the previous 5 years (31.9 versus 36.3 per 1,000).

TABLE 145

BIRTH DEFECT CASES, NSW 2000–2006#

Year	Birth defect cases	Births	Rate/1000 births
2000	1858	87140	21.3
2001	1775	85286	20.8
2002	1739	85398	20.4
2003	1761	85853	20.5
2004	1750	85016	20.6
2005	1621	89840	18.0
2006	792	92038	8.6

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

For 2000–2005, cases reported during pregnancy and up to one year of age are included. For 2006, cases reported during pregnancy or at birth are reported.

TABLE 146

BIRTH DEFECTS AMONG STILLBIRTHS AND LIVE BORN INFANTS BY DIAGNOSTIC CATEGORY, NSW 2000–2006#

Diagnostic category	No. defects				Rate/1,000 births			
	2000–2004	2005	2006	2000–2006	2000–2004	2005	2006	2000–2006
Defects of nervous system								
Anencephaly	51	10	7	68	0.1	0.1	0.1	0.1
Spina Bifida	128	25	18	171	0.3	0.3	0.2	0.3
Encephalocele	31	5	5	41	0.1	0.1	0.1	0.1
Microcephaly	117	16	6	139	0.3	0.2	0.1	0.2
Congenital hydrocephalus	180	26	20	226	0.4	0.3	0.2	0.4
Other nervous system defects	316	39	26	381	0.7	0.4	0.3	0.6
TOTAL	823	121	82	1026	1.9	1.3	0.9	1.7
Defects of eye								
Anophthalmos–microphthalmos	56	12	3	71	0.1	0.1	0.0	0.1
Buphthalmos–congenital glaucoma	25	9	1	35	0.1	0.1	0.0	0.1
Congenital cataract	67	21	3	91	0.2	0.2	0.0	0.1
Other eye defects	164	36	14	214	0.4	0.4	0.2	0.4
TOTAL	312	78	21	411	0.7	0.9	0.2	0.7
Defects of ear, face and neck								
Absence–stricture auditory canal	51	10	8	69	0.1	0.1	0.1	0.1
Absent auricle	8	2	–	10	0.0	0.0	.	0.0
Defects of face and neck	44	3	5	52	0.1	0.0	0.1	0.1
Other ear defects	79	8	14	101	0.2	0.1	0.2	0.2
TOTAL	182	23	27	232	0.4	0.3	0.3	0.4

TABLE 146

BIRTH DEFECTS AMONG STILLBIRTHS AND LIVE BORN INFANTS BY DIAGNOSTIC CATEGORY, NSW 2000–2006#

Diagnostic category	No. defects				Rate/1,000 births			
	2000–2004	2005	2006	2000–2006	2000–2004	2005	2006	2000–2006
Defects of cardiovascular system								
Transposition of great vessels	222	56	22	300	0.5	0.6	0.2	0.5
Tetralogy of Fallot	148	27	13	188	0.3	0.3	0.1	0.3
Ventricular septal defect	869	138	73	1080	2.0	1.5	0.8	1.8
Atrial septal defect	827	138	84	1049	1.9	1.5	0.9	1.7
Heart valve defects	552	111	46	709	1.3	1.2	0.5	1.2
Patent ductus arteriosus > 37 weeks	445	87	39	571	1.0	1.0	0.4	0.9
Coarctation of aorta	203	40	23	266	0.5	0.4	0.2	0.4
Other defects of aorta	101	25	17	143	0.2	0.3	0.2	0.2
Defects of pulmonary artery	141	32	19	192	0.3	0.4	0.2	0.3
Other cardiovascular defects	709	146	79	934	1.7	1.6	0.9	1.5
TOTAL	4217	800	415	5432	9.8	8.9	4.5	8.9
Defects of respiratory system								
Defects of nose	59	11	8	78	0.1	0.1	0.1	0.1
Defects of larynx, trachea and bronchus	49	6	1	56	0.1	0.1	0.0	0.1
Defects of lung	70	19	11	100	0.2	0.2	0.1	0.2
TOTAL	178	36	20	234	0.4	0.4	0.2	0.4
Defects of gastrointestinal system								
Cleft palate only	406	95	40	541	0.9	1.1	0.4	0.9
Cleft lip only	151	23	29	203	0.4	0.3	0.3	0.3
Cleft palate and cleft lip	223	34	38	295	0.5	0.4	0.4	0.5
Oesophageal atresia only	11	4	3	18	0.0	0.0	0.0	0.0
Tracheo-oesophageal fistula (TOF) only	19	5	4	28	0.0	0.1	0.0	0.0
Oesophageal atresia with TOF	70	17	15	102	0.2	0.2	0.2	0.2
Atresia/stenosis of small intestine	133	30	14	177	0.3	0.3	0.2	0.3
Atresia/stenosis of anus	115	26	18	159	0.3	0.3	0.2	0.3
Other gastrointestinal defects	464	86	30	580	1.1	1.0	0.3	0.9
TOTAL	1592	320	191	2103	3.7	3.6	2.1	3.4
Defects of genitourinary system								
Defects of female genitals	55	14	9	78	0.1	0.2	0.1	0.1
Undescended testis	305	36	16	357	0.7	0.4	0.2	0.6
Hypospadias	816	146	80	1042	1.9	1.6	0.9	1.7
Epispadias	11	.	3	14	0.0	.	0.0	0.0
Chordee	134	28	20	182	0.3	0.3	0.2	0.3
Indeterminate sex-ambiguous genitalia	54	12	9	75	0.1	0.1	0.1	0.1
Renal agenesis-dysgenesis	197	29	26	252	0.5	0.3	0.3	0.4
Obstructive defects of renal pelvis and ureter	703	118	36	857	1.6	1.3	0.4	1.4
Other genitourinary system defects	615	122	59	796	1.4	1.4	0.6	1.3
TOTAL	2890	505	258	3653	6.7	5.6	2.8	6.0
Defects of musculoskeletal system								
Congenital dislocation of the hips	682	140	28	850	1.6	1.6	0.3	1.4
Talipes equinovarus	344	73	26	443	0.8	0.8	0.3	0.7
Polydactyly	484	93	79	656	1.1	1.0	0.9	1.1
Syndactyly	86	25	18	129	0.2	0.3	0.2	0.2
Reduction deformities of limbs	226	50	49	325	0.5	0.6	0.5	0.5
Craniosynostosis	276	32	3	311	0.6	0.4	0.0	0.5
Diaphragmatic hernia	119	28	20	167	0.3	0.3	0.2	0.3
Exomphalos	74	11	9	94	0.2	0.1	0.1	0.2
Gastroschisis	90	17	20	127	0.2	0.2	0.2	0.2
Other musculoskeletal defects	784	137	106	1027	1.8	1.5	1.2	1.7
TOTAL	3165	606	358	4129	7.4	6.7	3.9	6.8
Defects of integumentary system								
Cystic hygroma	320	36	19	375	0.7	0.4	0.2	0.6
Chromosomal defects	50	13	3	66	0.1	0.1	0.0	0.1
Trisomy 21								
Trisomy 21	511	73	55	639	1.2	0.8	0.6	1.0
Trisomy 13								
Trisomy 13	36	6	7	49	0.1	0.1	0.1	0.1
Trisomy 18								
Trisomy 18	85	16	18	119	0.2	0.2	0.2	0.2
Turner syndrome								
Turner syndrome	70	14	7	91	0.2	0.2	0.1	0.1
Other chromosomal defects								
Other chromosomal defects	300	70	18	388	0.7	0.8	0.2	0.6
TOTAL	1002	179	105	1286	2.3	2.0	1.1	2.1
Situs inversus								
Situs inversus	27	5	1	33	0.1	0.1	0.0	0.1
Congenital malformation syndromes								
Congenital malformation syndromes	201	34	19	254	0.5	0.4	0.2	0.4
Congenital cytomegalovirus infection								
Congenital cytomegalovirus infection	2	0	0	2	0.0	0.0	0.0	0.0
Non-immune hydrops foetalis								
Non-immune hydrops foetalis	110	20	10	140	0.3	0.2	0.1	0.2
Other and unspecified birth defects								
Other and unspecified birth defects	491	93	15	599	1.1	1.0	0.2	1.0
TOTAL	15562	2869	1544	19975	36.3	31.9	16.8	32.7

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

For 2000–2005, cases reported during pregnancy and up to one year of age are included. For 2006, cases reported during pregnancy or at birth are reported.

Infant characteristics

In the period 2000–2006, a single defect was reported in 62.4 per cent of infants, 2 defects in 17.8 per cent, 3 defects in 8.3 per cent, and 4 or more defects in 11.6 per cent of cases. The sex was male in 57.9 per cent of infants, female in 41.6 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 147). Birth defects were also more common in infants born of a

multiple pregnancy than a singleton pregnancy: in 2000–2006, 1.8 per cent of singleton babies, 2.4 per cent of twins, and 3.6 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 148). 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 2006 (see Chapter 4).

TABLE 147

BIRTH DEFECT CASES BY GESTATIONAL AGE, NSW 2000–2006#

Gestational age (weeks)	2000–2004		2005		Year 2006		2002–2006		
	No.	%	No.	%	No.	%	No.	%	Rate/1,000 births
20 – 27	590	6.6	108	6.7	78	9.8	776	6.9	181.9
28 – 31	276	3.1	36	2.2	31	3.9	343	3.0	75.5
32 – 36	1062	12.0	230	14.2	114	14.4	1406	12.4	40.0
37 – 41	6355	71.5	1111	68.5	555	70.1	8021	71.0	14.5
42 +	183	2.1	25	1.5	13	1.6	221	2.0	16.6
Not stated	417	4.7	111	6.8	1	0.1	529	4.7	–
TOTAL	8883	100.0	1621	100.0	792	100.0	11296	100.0	18.5

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

For 2000–2005, cases reported during pregnancy and up to one year of age are included. For 2006, cases reported during pregnancy or at birth are reported.

TABLE 148

BIRTH DEFECT CASES BY PREGNANCY OUTCOME, NSW 2000–2006#

Pregnancy outcome	2000–2004		2005		Year 2006		2000–2006	
	No.	%	No.	%	No.	%	No.	%
Stillbirth	571	6.4	114	7.0	83	10.5	768	6.8
Liveborn–neonatal death	358	4.0	58	3.6	51	6.4	467	4.1
Liveborn–postneonatal death	68	0.8	9	0.6	1	0.1	78	0.7
Liveborn surviving	7886	88.8	1440	88.8	657	83.0	9983	88.4
TOTAL	8883	100.0	1621	100.0	792	100.0	11296	100.0

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

For 2000–2005, cases reported during pregnancy and up to one year of age are included. For 2006, cases reported during pregnancy or at birth are reported.

Maternal characteristics

After 35 years of age, the incidence of birth defects increased with increasing maternal age (Table 149). While the rate of birth defects is higher in older women, the majority of births occur in younger women. In 2000–2006, 72.1 per cent of babies with birth defects were born to women aged less than 35 years.

In 2000–2006, 330 babies of Aboriginal or Torres Strait Islander mothers were reported to have birth defects. The rate of birth defects among these babies was 20.6 per 1,000 compared with 17.0 per 1,000 for non-Aboriginal mothers.

TABLE 149

BIRTH DEFECT CASES BY MATERNAL AGE, NSW 2000–2006#

Maternal age (years)	Year								
	2000–2004		2005		2006		2000–2006		
	No.	%	No.	%	No.	%	No.	%	Rate/1,000
Under 20	361	4.1	62	3.8	39	4.9	462	4.1	18.5
20 – 24	1239	13.9	223	13.8	126	15.9	1588	14.1	17.8
25 – 29	2308	26.0	359	22.1	192	24.2	2859	25.3	16.4
30 – 34	2530	28.5	439	27.1	267	33.7	3236	28.6	16.0
35 – 39	1320	14.9	269	16.6	129	16.3	1718	15.2	17.3
40 – 44	320	3.6	78	4.8	36	4.5	434	3.8	22.4
45+	25	0.3	6	0.4	3	0.4	34	0.3	36.0
Not stated	780	8.8	185	11.4	0	0.0	965	8.5	–
TOTAL	8883	100.0	1621	100.0	792	100.0	11296	100.0	18.5

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

For 2000–2005, cases reported during pregnancy and up to one year of age are included. For 2006, 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 2000–2006, on average about 300 terminations of pregnancy per year were reported to the NSW Birth Defects Register (Table 150). To date, 196 terminations of pregnancy have been reported to the Register for 2006.

This number is expected to increase as outcomes for mothers with defects detected during pregnancy in 2006 continue to be reported. Of the 1,919 terminations of pregnancy reported in 2000–2006, 1,480 (77.1 per cent)

were associated with a chromosomal abnormality, the most common of which was Trisomy 21 (Down syndrome), and 192 (10.0 per cent) were associated with a neural tube defect (Tables 150 and 151).

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 150

PREGNANCIES WITH FETUSES AFFECTED BY BIRTH DEFECTS RESULTING IN SPONTANEOUS ABORTION, TERMINATION OF PREGNANCY OR UNKNOWN OUTCOME, NSW 2000–2006

Pregnancy outcome	Year							
	2000	2001	2002	2003	2004	2005	2006	2000–2006
	No.	No.	No.	No.	No.	No.	No.	No.
Spontaneous abortion	124	171	202	232	298	317	348	1692
Termination of pregnancy less than 20 weeks gestation	262	257	285	312	305	302	196	1919
Unknown outcome	22	19	7	18	38	29	0	133
TOTAL	408	447	494	562	641	648	544	3744

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

TABLE 151

BIRTH DEFECTS AMONG TERMINATIONS, SPONTANEOUS ABORTIONS AND UNKNOWN OUTCOMES OF PREGNANCY BY DIAGNOSTIC CATEGORY, NSW 2000–2006

Diagnostic category	Year						Termination of pregnancy less than 20 weeks gestation	Spont. abortion	Unknown	Spont. abortion	Termination of pregnancy less than 20 weeks gestation	
	2000–2004		2005		2006							2000–2006
	No.	Spont. abortion	Termination of pregnancy less than 20 weeks gestation	Unknown	Spont. abortion	Termination of pregnancy less than 20 weeks gestation	No.	No.	No.	No.	No.	
Defects of nervous system												
Neural tube defects	6		142	3	2	27	0	1	9	23	192	3
Other nervous system defects	6		111	5	2	25	0	1	9	30	166	5
TOTAL	12		253	8	4	52	0	2	18	53	358	8
Defects of eye	0		2	0	0	2	0	0	0	0	4	0
Defects of ear, face and neck	1		8	1	0	3	0	0	1	3	14	1
Defects of cardiovascular system	12		157	6	4	74	1	7	23	58	289	7
Defects of respiratory system	2		12	4	2	10	0	3	7	2	24	4
Defects of Gastrointestinal system	9		111	4	2	34	0	3	14	24	169	4
Defects of musculoskeletal system	28		255	10	3	69	1	3	34	63	387	11
Defects of Genitourinary system	9		133	4	2	19	1	1	12	51	203	5
Defects of the integumentary system	1		1	0	0	0	0	0	1	0	1	0
Cystic hygroma	12		108	5	4	21	2	4	20	14	143	7
Chromosomal defects												
Trisomy 21	79		565	34	29	138	14	24	132	69	772	48
Trisomy 13	39		80	13	17	19	1	15	71	6	105	14
Trisomy 18	52		188	5	11	25	7	9	72	20	233	12
Turner syndrome	106		99	8	40	17	0	41	187	11	127	8
Other chromosomal defects	735		194	30	218	34	4	252	1205	15	243	34
TOTAL	1011		1126	90	315	233	26	341	1667	121	1480	116
Situs inversus	0		4	0	0	1	0	0	0	0	5	0
Congenital Malformation syndromes	2		8	0	0	5	0	0	2	5	18	0
Non-immune hydrops foetalis	8		45	1	4	9	0	2	14	14	68	1
Other and unspecified birth defects	5		23	3	0	9	0	1	6	4	36	3
TOTAL	1112		2246	136	340	541	31	367	1819	412	3199	167

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 152 and Figures 14 to 17. For 2000–2005, malformations reported up to one year of age are included; for 2006, malformations reported during pregnancy or at birth are included.

The reported number of live born and stillborn infants with neural tube defects was 41 in 2000 and 40 in 2005, and 30 have been reported for 2006 to date. The number of reported terminations of pregnancy was 36 in 2000, 26 in 2005, and 22 in 2006 (Figure 14).

Over the period 2000–2006, the number of cases of isolated cleft palate ranged from 60 to 90 per year, and for total cleft lip (including cases of cleft lip and cleft palate) from 66 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 live born and stillborn infants with chromosomal defects was 219 in 2000 and 179 in 2005. The number of reported terminations of pregnancy associated with chromosomal defects rose from 193 in 2000 to 233 in 2005. The number of infants born with Down syndrome was 118 in 2000 and 73 in 2005, while the number of reported terminations of pregnancy associated with Down syndrome rose from 96 in 2000 to 138 in 2005 (Figures 15 and 16).

In 2000, 20 live born infants were reported to have a diaphragmatic hernia, and there were 2 terminations of pregnancy associated with this condition. In 2005, there were 28 live born infants reported with a diaphragmatic hernia, and there were no terminations of pregnancy (Figure 17).

TABLE 152

SELECTED BIRTH DEFECTS BY YEAR, NSW 2000–2006#

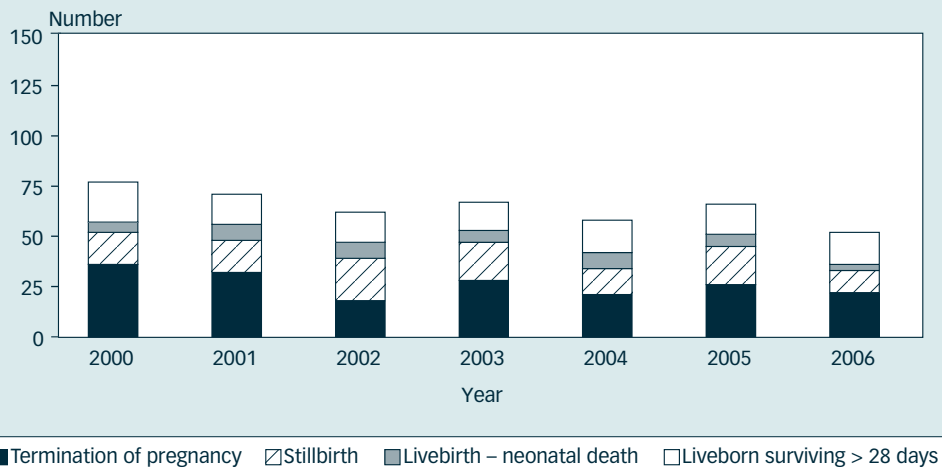
Birth defect	2000		2001		2002		2003		2004		2005		2006	
	No.	Rate/ 1000	No.	Rate/ 1000	No.	Rate/ 1000	No.	Rate/ 1000	No.	Rate/ 1000	No.	Rate/ 1000	No.	Rate/ 1000
Neural tube defects	77	0.9	71	0.8	62	0.7	67	0.8	58	0.7	66	0.73464	52	0.6
Anencephalus	29	0.3	25	0.3	20	0.2	18	0.2	16	0.2	19	0.21149	13	0.1
Spina bifida	42	0.5	39	0.5	33	0.4	46	0.5	35	0.4	40	0.44524	32	0.3
Encephalocele	13	0.1	8	0.1	10	0.1	7	0.1	8	0.1	8	0.08905	8	0.1
Cleft palate	79	0.9	67	0.8	60	0.7	88	1.0	74	0.9	90	1.00178	39	0.4
Total cleft lip	71	0.8	88	1.0	76	0.9	85	1.0	79	0.9	66	0.73464	69	0.7
Hypospadias	191	2.2	173	2.0	133	1.6	171	2.0	150	1.8	146	1.62511	80	0.9
Limb reduction defects	61	0.7	42	0.5	21	0.2	31	0.4	36	0.4	38	0.42297	39	0.4
Chromosomal abnormalities	412	4.7	370	4.3	441	5.2	451	5.3	446	5.2	412	4.58593	226	2.5
Down syndrome	214	2.5	180	2.1	221	2.6	227	2.6	232	2.7	211	2.34862	124	1.3
Renal agenesis and dysgenesis	82	0.9	75	0.9	63	0.7	68	0.8	82	1.0	73	0.81256	58	0.6
Exomphalos	28	0.3	22	0.3	22	0.3	20	0.2	28	0.3	15	0.16696	19	0.2
Gastroschisis	20	0.2	23	0.3	18	0.2	19	0.2	16	0.2	19	0.21149	21	0.2
Diaphragmatic hernia	22	0.3	28	0.3	23	0.3	21	0.2	30	0.4	28	0.31167	21	0.2

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

For 2000–2005, cases reported during pregnancy and up to one year of age are included. For 2006, cases reported during pregnancy or at birth are reported.

FIGURE 14

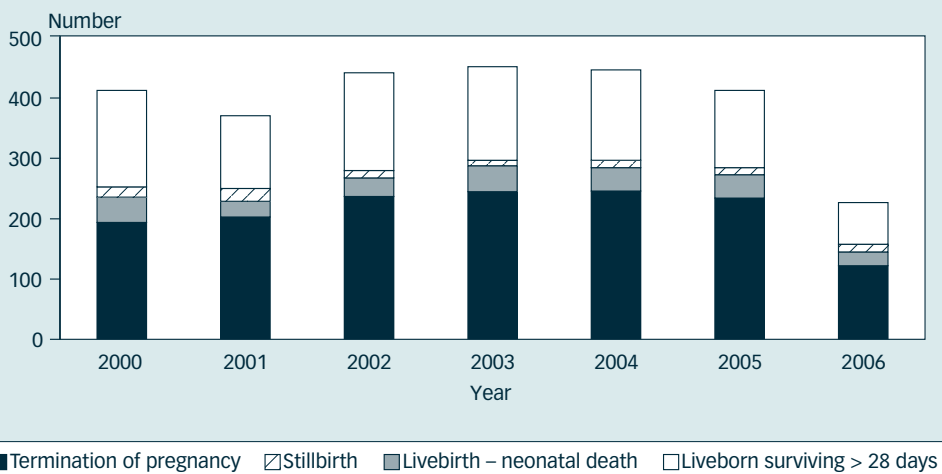
NEURAL TUBE DEFECTS: CASES BY YEAR AND PREGNANCY OUTCOME, NSW 2000–2006



Source: NSW Birth Defects Register. Centre for Epidemiology and Research. NSW Department of Health.
For 2000–2005, cases reported during pregnancy and up to one year of age are included. For 2006, cases reported during pregnancy or at birth are reported.

FIGURE 15

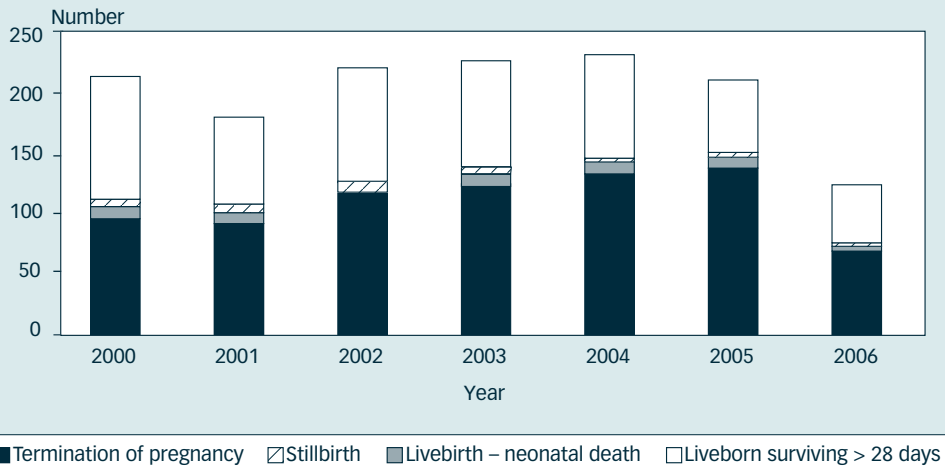
CHROMOSOMAL ABNORMALITIES: CASES BY YEAR AND PREGNANCY OUTCOME, NSW 2000–2006



Source: NSW Birth Defects Register. Centre for Epidemiology and Research. NSW Department of Health.
For 2000–2005, cases reported during pregnancy and up to one year of age are included. For 2006, cases reported during pregnancy or at birth are reported.

FIGURE 16

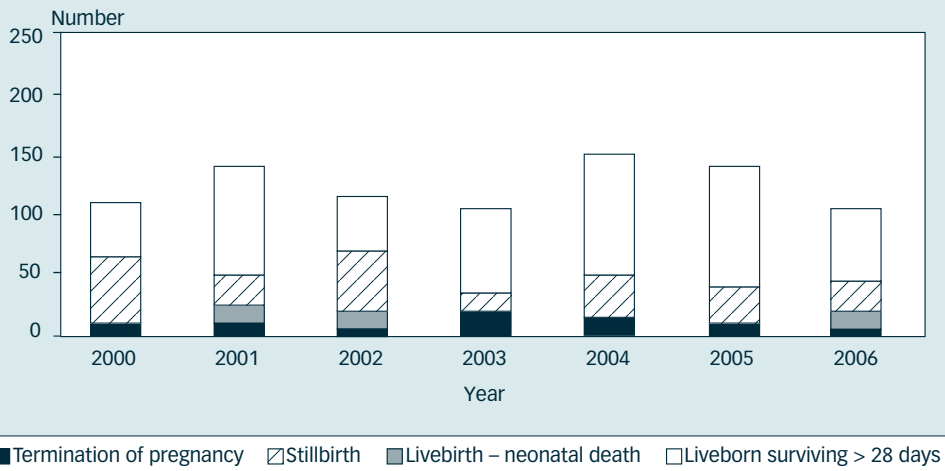
DOWN SYNDROME: CASES BY YEAR AND PREGNANCY OUTCOME, NSW 2000–2006



Source: NSW Birth Defects Register. Centre for Epidemiology and Research. NSW Department of Health.
 # For 2000–2005, cases reported during pregnancy and up to one year of age are included. For 2006, cases reported during pregnancy or at birth are reported.

FIGURE 17

DIAPHRAGMATIC HERNIA CASES BY YEAR AND PREGNANCY OUTCOME, NSW 2000–2006*



Source: NSW Birth Defects Register. Centre for Epidemiology and Research. NSW Department of Health.
 # For 2000–2005, cases reported during pregnancy and up to one year of age are included. For 2006, 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 153. The denominator population includes live births and stillbirths among NSW residents as reported to the MDC. The rate of birth defects increases with increasing maternal age (Table 149). 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 2000–2006, standardised rates of reported birth defects were lowest in the Greater Southern Area and highest in the Hunter and New England Area. Review of cases showed slightly increased reported rates of a range of birth defects in the Hunter and New England Area compared to NSW overall including: unstable hips (but not dislocated hips), first degree hypospadias, undescended testis, obstructive defects of the renal pelvis and ureter, spina bifida, ventricular septal defect, atrial septal defect and heart valve defects. There was no clustering by small geographic area. The range and pattern of these defects suggests that reporting of congenital abnormalities is better in the Hunter and New England Health Area compared with NSW as a whole.

TABLE 153

BIRTH DEFECTS IN NSW HEALTH AREAS, 2000–2006#

Health Area	2000–2004			2005			2006			2000–2006			99% confidence interval
	No. of cases	Crude rate per 1000 births	Standardised rate per 1000 births	No. of cases	Crude rate per 1000 births	Standardised rate per 1000 births	No. of cases	Crude rate per 1000 births	Standardised rate per 1000 births	No. of cases	Crude rate per 1000 births	Standardised rate per 1000 births	
Sydney South West	2270	23.4	21.0	373	18.6	16.3	202	9.7	8.5	2845	20.7	18.5	17.5–19.5
South Eastern Sydney & Illawarra	1703	24.1	20.8	314	21.1	18.9	174	11.4	11.2	2191	21.7	19.1	17.8–20.4
Sydney West	1840	22.8	20.9	297	17.2	15.0	172	9.8	9.6	2309	20.0	18.3	17.2–19.4
Northern Sydney & Central Coast	1537	23.2	19.4	309	22.2	15.6	161	11.2	9.8	2007	21.2	17.4	16.1–18.8
Hunter & New England	1401	28.0	25.4	302	28.6	25.6	137	12.8	11.5	1840	25.8	23.4	21.9–24.9
North Coast	469	19.8	18.8	90	17.9	16.4	49	9.5	8.3	608	17.9	16.9	15.1–18.8
Greater Southern	375	18.4	16.5	83	20.5	15.3	31	7.5	8.0	489	17.1	15.1	13.3–17.1
Greater Western	424	21.1	19.8	82	20.3	18.8	57	13.9	12.9	563	19.9	18.7	16.6–20.9
TOTAL NSW	10019	23.4	20.9	1850	20.6	17.7	983	10.7	9.9	12852	21.0	18.8	18.3–19.3

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

For 2000–2005, cases reported during pregnancy and up to one year of age are included. For 2006, cases reported during pregnancy or at birth are reported.