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Neural tube defects in Australia

Prevalence before mandatory folic acid fortification

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Abbreviations

ABS	Australian Bureau of Statistics
ACAMS	Australian Congenital Anomalies Monitoring System
ACT	Australian Capital Territory
AHMAC	Australian Health Ministers' Advisory Council
AIHW	Australian Institute of Health and Welfare
ASGC	Australian Statistical Geographical Classification
BPA	British Paediatric Association
CI	confidence interval
FSAG	Folate Scientific Advisory Group
FSANZ	Food Standards Australia New Zealand
ICD-9-BPA	International Classification of Diseases, 9th Revision, British Paediatric Association Publication
ICD-10-AM	International Statistical Classification of Diseases and Related Health Problems, 10th Revision, Australian Modification
LB	live births
NHMRC	National Health and Medical Research Council
NPDC	National Perinatal Data Collection
NPESU	National Perinatal Epidemiology and Statistics Unit
NSW	New South Wales
NT	Northern Territory
NTD	neural tube defect
Qld	Queensland
SA	South Australia
SACC	Standard Australian Classification of Countries
SB	stillbirths
SEIFA	Socio-Economic Indexes for Areas
Tas	Tasmania
TB	total births
TOP	terminations of pregnancy
Vic	Victoria
WA	Western Australia

Symbols

–	nil or rounded to zero
..	not applicable
n.a.	not available
n.p.	not publishable because of small numbers, confidentiality or other concerns about the quality of the data
g	gram
µg	microgram

Summary

Background

Neural tube defects (NTDs) are a group of major congenital anomalies comprising anencephaly, spina bifida and encephalocele that result from very early disruption in the development of the brain and spinal cord. These conditions are often incompatible with life. Survivors frequently require intensive, costly, lifelong health and social care.

There is strong evidence of substantial reductions in the prevalence of NTD-affected pregnancies among women given periconceptional folic acid. Information about the benefits and programs promoting dietary and/or supplemental folic acid intake by women before and during pregnancy have existed in Australia for over 20 years. Voluntary fortification of foods with folic acid has been permitted in Australia since 1995. These measures have resulted in a very modest increase in folic acid consumption overall by women of childbearing age.

In Australia, from 13 September 2009, the mandatory folic acid fortification standard requires the addition of folic acid to all wheat flour for bread making, with the exception of organic bread, within the prescribed range of 200–300 μg per 100 g of flour. The main benefit is the primary prevention of NTDs.

This report updates the baseline prevalence of NTDs in Australia using data for the years from 2006 to 2008. These precede the implementation of mandatory folic acid fortification of bread flour. It is expected that the information provided in this report will help evaluate the impact of mandatory folic acid fortification on the prevalence of NTDs in the future.

Key findings

The results demonstrate four key aspects of the overall prevalence of NTDs that have implications for monitoring the effects of bread flour fortification.

- A continued downward trend was observed in the overall prevalence of all NTDs. Between 1998 and 2008 the average annual decline in the overall prevalence of NTDs was 0.22 NTD per 10,000 births per year.
- There were different trends in the overall prevalence of individual NTDs, i.e. anencephaly, spina bifida and encephalocele. Between 1998 and 2008 there was a fall in the overall prevalence of spina bifida, but no appreciable change in the overall prevalence of anencephaly or encephalocele.
- There was unequal distribution of the NTD overall prevalence across the population with higher prevalence of NTD-affected pregnancies at extremes of reproductive age, that is, among women in the youngest and oldest age groups; among women living in areas of relative disadvantage; among women living in remote areas; and among women of Aboriginal and Torres Strait Islander origin.
- Reduced overall prevalence of NTDs does not necessarily reduce birth prevalence of NTDs. Rising birth prevalence of NTDs was seen between 2005 and 2008 in the context of lower overall prevalence of NTDs in Victoria and there was no evidence of change in the overall prevalence of NTDs in New South Wales.

1 Background

1.1 What are neural tube defects?

Neural tube defects (NTDs) are congenital anomalies affecting approximately one in every thousand pregnancies. NTDs result from the very early disruption of the development of the brain and/or the spinal cord. The first stage in the development of these organs is the formation of the neural tube. If this does not close along its entire length it causes disruption to the proper development of the nerve cells and the supporting tissues that cover them. The neural tube has normally closed completely by the end of the fourth week after fertilisation.

There are three distinct forms of NTDs depending on the part of the neural tube affected. The brain is formed by the upper part of the neural tube and failed closure of this part results in anencephaly or encephalocele. Spina bifida defects result from defects in closure of a lower part of the neural tube that supports the formation of the spinal cord.

The consequences of NTDs vary according to their extent and site. Anencephaly is not usually compatible with life outside the womb. Babies with anencephaly born alive rarely survive for more than a few days. Babies born with spina bifida or encephalocele survive unless the condition is severe. The site and extent of brain tissue involved in encephalocele will determine the extent of physical and intellectual impairment. With spina bifida any part of the body that receives its nerve supply from the spinal level at or below the defect can be affected. Lower limb paralysis and incontinence are therefore common. Additional fluid around the brain (hydrocephalus) can also be present and cause additional brain damage.

Babies born with spina bifida and encephalocele may need neonatal intensive care, surgery, rehabilitation and other specialised management. Surviving infants with spina bifida are likely to have lifelong impairment and disabilities (Date et al. 1993), which may include learning difficulties. While some survivors will have profound and multiple disabilities, in others impairment or disability will be minimal. Overall the lifetime costs of caring for a child with a NTD can be substantial.

There may be more than one NTD present in one baby. They may occur in isolation, or together with other congenital anomalies. Some anomalies, such as talipes equinovarus (a deformity seen in the foot) are considered to be the consequence of the NTD rather than being a separate anomaly.

1.2 What causes neural tube defects?

The effort to understand NTDs over the past 30 years has generated much literature, but the mechanisms underlying the development of NTDs in humans remain poorly understood. Most cases of NTD are of multi-factorial origin involving interactions between genes and the environment. A review by Padmanabhan (2006) is summarised in the three paragraphs below.

NTDs aggregate in some families. There is a 2–5% risk of NTD recurrence in subsequent pregnancies. NTD occurs more often in near relatives of an NTD-affected baby than in the population as a whole, but less than the 25% or 50% expected from simple dominant or recessive gene inheritance. NTDs, particularly anencephaly, occur more frequently among twins than singletons. Twinning is increased among relatives of babies with anencephaly,

and such families have higher rates of NTD recurrence than families without twins. Nutrition plays a key role in the development of NTDs. Behavioural and genetic factors singly and together can affect the supply of critical nutrients. Nutritional deficiencies involving several vitamins and trace elements have been implicated. The best studied is the folate group.

A very small proportion of NTDs occur as part of known genetic syndromes or chromosomal abnormalities such as trisomy 13, trisomy 18 and certain chromosome rearrangements.

The majority of cases occur in families with no history of NTD. Environmental agents, such as hyperthermia from the use of saunas, hot tub and maternal pyrexia, at critical periods of development may play a role in the development of some NTDs. Exposures to other noxious agents such as organic solvents, anaesthetic agents, pesticides and X-radiation have been reported to increase the risk of NTDs.

1.3 The role of folate in neural tube defect prevention

Folate is a B group vitamin necessary for the production and maintenance of new cells, which is especially important during periods of rapid cell division and growth. Initial observations that linked the occurrence of NTDs with folate deficiencies were tested in the 1980s with randomised controlled trials of the use of vitamin supplementation with folic acid (the synthetic form of folate) to prevent recurrence of NTDs. The first meta-analyses of these trials published in 1995 showed substantial reductions in NTD-affected pregnancies among subjects given periconceptional folic acid. These studies were combined and updated in the current Cochrane review (Lumley et al. 2001).

In Australia these findings led to a variety of public health programs in the 1990s aimed at increasing the use of folic acid among women of reproductive age. A more comprehensive discussion of the programs is presented in the 2008 report (Abeywardana & Sullivan 2008b). Broadly, these consisted of campaigns to increase awareness of the benefits of folic acid, and the promotion and/or provision of vitamin supplementation for women planning pregnancy. Such programs were recognised to be of limited value in populations not amenable to health education and where many pregnancies are unplanned. In 1994, the National Health and Medical Research Council (NHMRC) recommended fortification of food with folic acid (NHMRC 1995).

1.4 Fortification to prevent neural tube defects

Voluntary food fortification

Food manufacturers can add vitamins and minerals to food where there is evidence of a potential benefit and also that fortification will not result in harm. Food Standards Australia New Zealand (FSANZ) regulates the addition of vitamins and minerals to food. The voluntary addition of folic acid to certain foodstuffs has been permitted in Australia since 1996 (Abraham & Webb 2001). Since then a variety of products have been fortified with folic acid.

In the period following voluntary fortification, during which health education efforts continued, there was a reduction of NTDs in Australia (Chan et al. 2001; Halliday & Riley

2000; Bower et al. 2002). However, this reduction was not seen among all ethnic and socioeconomic groups. The interim evaluation of the voluntary folate fortification policy by Abraham and Webb (2001) collated results of studies carried out between 1995 and 1998 to conclude that the criterion for effectiveness defined by the NHMRC Expert Panel (at least 70% of women consuming more than 400 μg of folate per day) was not met under voluntary fortification. By November 1998, voluntary fortification had resulted in only a small increase in mean folate intake (11%) among the target population.

These findings, together with the overwhelming evidence from Australian and international research studies, led the way for the Australia and New Zealand Food Regulation Ministerial Council in May 2004 to request FSANZ to consider the mandatory fortification of food with folic acid.

Mandatory folic acid fortification

Mandatory fortification with folic acid was implemented in Australia from 13 September 2009. The mandatory fortification standard requires the addition of folic acid to all wheat flour for bread making, with the exception of organic bread making flour, within the prescribed range of 200–300 μg per 100 grams of flour (FSANZ 2007). This level of fortification is expected to prevent between 14 and 49 NTD per year in Australia, when combined with existing voluntary fortification permissions and current levels of supplemental use (FSANZ 2006).

In developing the mandatory folic acid fortification standard, FSANZ convened the Folate Scientific Advisory Group (FSAG), comprising clinicians and public health nutritionists with expertise in epidemiology and/or folate nutrition. This group comprehensively assessed the potential benefits and risks of increasing folic acid intake within the population (FSANZ 2006). Their findings are summarised below:

- The primary benefit of increased folate consumption is reduction in the prevalence of NTDs, which was estimated to fall between 40% and 70%. Mandatory fortification of bread flour with folic acid allows all women who become pregnant to have increased blood folate levels. Women who are not aware of the importance of folic acid, have unplanned pregnancies or are unable to afford supplements will benefit from increased folate levels resulting from folic acid fortification.
- A major concern about folic acid fortification was the potential to mask vitamin B12 deficiency, particularly in the elderly. At the level of intake from mandatory folic acid fortification there is no evidence of this, particularly as the diagnosis of vitamin B12 deficiency now relies on a combination of different clinical tests (FSANZ 2009). Further, a study in NSW (Flood et al. 2001) has shown that fortification at the levels proposed is unlikely to substantially increase the number of older people consuming folic acid over the safety limit. No conclusive evidence was found for other potential risks to health in the population.
- Bread flour was chosen for mandatory fortification because bread and bread products are staple foods that are consumed widely, consistently, and regularly by the target population of women aged 16–44 years. This strategy had already been applied in other countries with positive effects in reducing NTDs and no evidence of harm in the population as a whole.

1.5 Effects of folic acid fortification in other countries

The United States was the first country, commencing in 1997, to mandate the fortification of wheat flour with folic acid. Since then more than 50 countries have implemented mandatory fortification programs or regulations (Cordero et al. 2010). Decreased prevalence of NTDs has since been documented in several countries that have introduced mandatory folic acid fortification programs, such as the United States, Canada, Costa Rica, Chile and South Africa (Berry et al. 2010). The level of folic acid fortification and baseline prevalence of NTDs varies between countries and hence there are differences in prevalence before and after fortification was introduced.

The most recent reports of NTD prevalence in relation to folic acid fortification of flour in Canada (De Wals et al. 2007) and the United States (National Birth Defects Prevention Network 2010) are presented in Table 1.1.

Table 1.1: Changes in prevalence of NTDs in Canada and the United States after fortification with folic acid, 1993–2002

	Period			Change in NTD prevalence between periods of:			
	No fortification	Voluntary fortification	Mandatory fortification	voluntary and mandatory fortification		no and mandatory fortification	
Canada[a]	**Jan 1993 –Sept 1997**	**Oct 1997 –Mar 2000**	**Apr 2000 –Dec 2002**				
	Overall prevalence[c]			**Diff.**	**% diff.**	**Diff.**	**% diff.**
Anencephaly	5.2	3.8	3.2	0.6	15.8	2.0	38.5
Spina bifida	8.6	5.7	4.0	1.7	29.8	4.6	53.5
Encephalocele	1.7	1.2	1.2	—	—	0.5	29.4
All NTD	15.8	10.9	8.6	2.3	21.1	7.2	45.6
United States[b]	**Jan 1995 –Dec1996**	**Jan 1997 –Sept 1998**	**Oct 1998 –Dec 2006**				
	Overall prevalence[c]			**Diff.**	**% diff.**	**Diff.**	**% diff.**
Anencephaly	4.1	3.4	3.0	0.4	11.3	1.2	28.0
Spina bifida	6.6	5.5	4.0	1.5	27.7	2.7	40.5
	Birth prevalence[c]						
Anencephaly	2.1	2.0	1.6	0.4	21.3	0.6	26.9
Spina bifida	4.6	4.0	3.4	0.6	14.6	1.2	25.5

(a) De Wals et al. 2007.

(b) National Birth Defects Prevention Network 2010.

(c) Overall prevalence refers to estimates from data that include births and terminations of pregnancy due to congenital anomalies. Birth prevalence refers to estimates obtained from birth data only. Prevalence is reported per 10,000 total births.

Notes

1. Diff. is the absolute difference in NTD prevalence between specified time periods.
2. The % diff. is the absolute difference as a percentage of the prevalence during the first of the two time periods being compared.

The results demonstrate the absolute and percentage reduction in prevalence of NTDs between periods of no fortification and fortification, and between periods of voluntary and mandatory fortification. In Canada for example, when comparing NTD prevalence during the period of voluntary flour fortification and mandatory fortification there was an overall reduction of 2.3 NTDs per 10,000 total births, which represents a 21% reduction in overall prevalence of NTDs. The difference in overall prevalence of NTDs in Canada between the period in which there was no fortification and the period of mandatory folic acid fortification was 7.2 NTD per 10,000 total births. The overall prevalence of NTDs in Canada during mandatory fortification was 46% of the overall prevalence observed prior to any fortification. A similar reduction in overall prevalence in time periods characterised by different folic acid fortification standards was also evident in the United States.

1.6 Rationale for this report

This report updates the baseline prevalence of NTDs in Australia using data from 2006 to 2008. These precede the implementation of mandatory folic acid fortification of bread flour. It is expected that the information provided in this report will help evaluate the impact of mandatory folic acid fortification on the prevalence of NTDs in the future.

2 Methods

2.1 Definitions

Neural tube defect

Neural tube defect is the collective name for congenital anomalies that comprise anencephaly, spina bifida and encephalocele. Listed below are the simple definitions of these anomalies including the specific ICD-10-AM and ICD-9-BPA codes used to select records from congenital anomaly registers. The full definitions are set out in Appendix 1.

Anencephaly

The total or partial absence of the upper part of the brain, the bones that make up the top of the skull, and the skin that would cover these parts. The remaining brain tissue may be disorganised or damaged. The following codes were used to select records with this condition:

ICD-9-BPA codes: 740.00–740.29 or ICD-10-AM codes: Q00.0–Q00.2.

Spina bifida

An opening in the skin and one or more of the backbones (the spinal column) that exposes the spinal cord, nerves or the tissues that covers them. The exposed nerves and spinal cord may be disorganised or damaged. The following codes were used to select records with this condition:

ICD-9-BPA codes: 741.00–741.99 or ICD-10-AM codes: Q05.0–Q05.9.

Encephalocele

An opening in the skull bones that exposes part of the brain or the tissues that covers it. The following codes were used to select records with this condition:

ICD-9-BPA codes: 742.00–742.09 or ICD-10-AM codes: Q01.0–Q01.2, Q01.8, Q01.9.

Total births

Total births include both stillbirths and live births of babies of at least 20 weeks gestational age or 400 grams birthweight. This definition is the same as that used for a birth by the National Perinatal Data Collection (NPDC).

Overall prevalence

A ratio with the numerator defined as the number of NTDs among pregnancies that ended in a birth or a termination of pregnancy at any gestation for congenital anomaly in a defined population and the denominator defined as the number of total births in that population.

Birth prevalence

The birth prevalence is the proportion of NTD affected births among total births in a defined population.

2.2 Data sources

Australian Congenital Anomalies Monitoring System

The Australian Congenital Anomalies Monitoring System (ACAMS) collates state and territory data about structural and chromosomal anomalies from pregnancies ending either in a birth (defined as a live birth or a stillbirth of at least 20 weeks gestational age or 400 grams birthweight) or induced abortions carried out before 20 weeks gestational age and notified to the state or territory birth defect register or perinatal data collection. Information about congenital anomalies is collected in all Australian states and territories except the Northern Territory. Descriptions of the jurisdictional collections are set out in Appendix 2.

The range of data requested for ACAMS is listed in Appendix 3 and includes details about the affected baby or fetus, the woman who was pregnant and the pregnancy. Detailed descriptions of each of the data items used in this report are set out in Appendix 4.

Data obtained for this report

Information about NTD notified to a state or territory congenital anomaly register or perinatal data collection between 2006 and 2008 was requested for this report. The data were added to ACAMS data for the years between 1998 and 2005 which were used for the previously published *Neural tube defects in Australia* (Abeywardana & Sullivan 2008b) and *Congenital anomalies in Australia* (Abeywardana & Sullivan 2008a). Data was supplied by six of the eight jurisdictions. Northern Territory does not collect congenital anomaly data and Tasmania, for logistic reasons, could not supply data for this report. There was a change in the criteria for data supplied by Queensland. For the years 1998–2005 Queensland provided data for NTD-affected births that were at least 20 weeks gestation **and** 400 grams birthweight. From 2006 to 2008, Queensland provided data on NTD-affected births that were at least 20 weeks gestation **or** 400 grams birthweight.

The numbers of total births were obtained from the NPDC as tables stratified by the relevant time periods and maternal, pregnancy or baby characteristics.

Measures used in this report

Calculating NTD prevalence

Two measures are used in this report. The overall prevalence measures NTDs among pregnancies that ended in a birth or a termination of pregnancy for congenital anomaly at any gestation in a defined population. The birth prevalence measures NTDs among live births and stillbirths in a defined population. The denominator for both prevalence measures, in line with international convention (ICBDSR 2008), is the number of total births in the population. While birth prevalence is a proportion, overall prevalence is, strictly speaking, a ratio because the numerator is not a subset of the denominator. The overall prevalence closely approximates the proportion of births and terminations for a congenital anomaly as the number of terminations of pregnancy is very small compared with the number of births. Both measures are expressed per 10,000 total births.

Why measure prevalence?

The incidence of NTD, which is the frequency with which the condition occurs in a defined population in a specified period of time, cannot be measured. This is because it is not possible to ascertain NTD-affected pregnancies that end with a spontaneous miscarriage or therapeutic terminations of pregnancy for reasons other than a diagnosis of congenital anomaly, and hence obtain a measure of the actual number of occurrences. Products of conceptions from women who receive care for spontaneous miscarriage or induced abortion are not systematically examined. Many early miscarriages may not even be recognised. Research undertaken before ultrasound technologies were widely available examined the products of conception from spontaneously aborted pregnancies and NTDs among births. The researchers estimated that half of the NTD-affected fetuses at 8 weeks of gestation were lost before birth (Creasy & Alberman 1976). The usual measure of NTD, as with most congenital anomalies, is therefore to measure their prevalence among births and terminations of pregnancy with a diagnosis of a congenital anomaly (ICBDSR 2008).

Hierarchical classification of neural tube defects

A baby may be affected by more than one type of NTD. In line with the previous report (Abeywardana & Sullivan 2008b) and with international conventions (ICBDSR 2008), in situations where it is necessary to consider the prevalence of anencephaly, spina bifida and encephalocele separately but count each baby once only, a hierarchical classification is applied. In this classification the anencephaly group includes babies with spina bifida and encephalocele in addition to anencephaly, and the spina bifida group will include babies with both spina bifida and encephalocele. The encephalocele group includes babies with encephalocele only.

Neural tube defect - related conditions and unrelated conditions

Conditions other than those defined as neural tube conditions were listed and reviewed by a member of the expert group and classified as an NTD-related condition, an unrelated condition, or a condition that required further information before it could be classified as a related or an unrelated condition. The list of conditions and their classification is provided in Appendix 4.

Mortality ratio

When considering the mortality of babies with NTD it is necessary to include fetal deaths from pregnancies terminated for congenital anomaly before 20 weeks gestational age in addition to stillbirths and neonatal deaths. The NTD mortality ratio relates the number of these deaths, which form the numerator, to the denominator, which is the number of total births (live births and stillbirths) in the corresponding period. This ratio closely approximates the NTD fetal mortality rate, which includes the currently unavailable total number of pregnancies terminated before 20 weeks gestational age, in the denominator.

Geographic methods

Postcodes of mothers' usual place of residence provided by NSW, Victoria, South Australia and Western Australia were used to assign the postal area scores for Socio-Economic Indexes for Areas (SEIFA) and remoteness of area of usual residence to records of NTD-affected pregnancies from ACAMS. These were similarly assigned to births from the NPDC. The 2001 SEIFA Index of Disadvantage (ABS 2001b) was assigned to each record with a pregnancy

ending from 2000 to 2002 and the 2006 index (ABS 2008) was used to assign a score to records with pregnancies ending in and 1 year either side of the 2006 census year, that is, between 2005 and 2007. A SEIFA score could not be assigned for records without a valid postal code.

Postal areas were divided into one of four population quartiles according to the level of disadvantage with equal numbers of households, that is, the first quartile contains the 25% of the population that reside in the most disadvantaged areas, while the fourth quartile contains the 25% of the population that reside in the least disadvantaged areas. Quartile cut-off scores (ABS 2004) were used to assign records from between 2000 and 2002 the relevant quartile. SEIFA scores for the 25th, 50th and 75th percentiles (ABS 2008) were used to similarly assign the 2005 to 2007 records.

The remoteness of area of usual residence classification was assigned directly using the Australian Standard Geographical Classification (ASGC) 2001 (ABS 2001a) for years between 2000 and 2003 (ABS 2004) and ASGC 2006 for from 2005 to 2008 (ABS 2008). In this report, the remoteness areas of usual residence are presented under three headings: Major cities, Regional (includes inner and outer regional) and Remote (includes remote and very remote).

The Standard Australian Classification of Countries (SACC) 2008 was used to assign a country of birth for mothers where a country of birth code was provided by the states and territories.

Statistical analysis

Precision of the estimates has been assessed by confidence intervals (CI) around prevalence measures, using the Poisson method to calculate the standard errors. The statistical significance of differences between prevalence measures was assessed using binomial methods to calculate standard errors that are applied to a standard normal distribution (Armitage & Berry 1994). Linear regression methods were used to calculate the line of best fit using the least squares method and test linear trend (Sokal & Rohlf 1995).

3 Results

3.1 Neural tube defect cases from states and territories

Six of the eight Australian states and territories provided information about NTD among births and four also provided information about pregnancies that ended with a termination of pregnancy for congenital anomaly before 20 weeks gestation. The Australian Capital Territory provided fewer than five cases in total from 2006 to 2008. These cases, which were not included in this report, were ascertained from hospital data only, a change from previous years when NTDs were ascertained from both perinatal and hospital data. Tasmania was unable to provide data for pregnancies ending between 2006 and 2008. Northern Territory does not currently collect information about congenital anomalies.

There were 729 reported cases of NTD affected pregnancies that ended as live births, stillbirths or terminations of pregnancy in the 3 years from 2006 to 2008 reported from Victoria, Western Australia, South Australia, New South Wales and Queensland. These were amalgamated with the 1,809 NTD babies from pregnancies ending in these states from 1998 to 2005 reported previously (Abeywardana & Sullivan 2008b). Table 3.1 sets out the numbers of NTD cases, the percentage distribution and prevalence of NTDs in these two time periods.

Between 2006 and 2008 the overall prevalence of NTDs in Victoria, Western Australia and South Australia was similar with 11.5, 12.4 and 12.1 NTD-affected pregnancies per 10,000 total births respectively.

The overall prevalence of 7.2 per 10,000 total births between 2006 and 2008 for births in New South Wales remains substantially lower than the overall prevalence in Victoria, Western Australia and South Australia over the same period. The shortfall is most evident in notifications of NTDs among pregnancies that ended in a termination of pregnancy. In New South Wales termination of pregnancy contributed 3.3 NTDs per 10,000 total births to the overall prevalence in 1998 to 2005 and 2.3 per 10,000 total births in 2006 to 2008. In Victoria, Western Australia and South Australia, termination of pregnancy contributed between 5.1 and 9.8 NTDs for every 10,000 births. The New South Wales Register of Congenital Conditions advises that congenital anomalies associated with terminations are likely to be under-reported.

From 2006 to 2008 the birth prevalence in New South Wales, Victoria, Western Australia and South Australia ranged from 2.6 NTD-affected pregnancies per 10,000 total births in Western Australia to 6.5 NTD-affected pregnancies per 10,000 births in Victoria. NTD reported among terminations of pregnancy for congenital anomaly before 20 weeks gestational age and weighing less than 400 grams at birth accounted for the remaining overall prevalence. The same pattern of results are seen when the percentage distributions of NTDs are compared across these jurisdictions.

In Queensland all NTD cases are ascertained from birth data. The higher birth prevalence between 2006 and 2008 in Queensland is partly due to different criteria applied to data provided by Queensland for NTD-affected births from 1998 to 2005 and 2006 to 2008. From 1998 to 2005 data for all NTD-affected births that were at least 20 weeks gestation and 400 grams were supplied. Between 2006 and 2008 data was supplied for all NTD-affected births

with at least 20 weeks gestation or 400 grams birthweight. This yielded an additional 36 stillbirths and 9 live births.

Table 3.1: Neural tube defects provided by state or territory, 1998–2005 and 2006–2008

	1998–2005					2006–2008[a]				
	Termi n-ations (TOP)	Still-births (SB)	Live births (LB)	Total births (TB= SB+LB)	All preg-nancies (TB+TOP)	Termi n-ations (TOP)	Still-births (SB)	Live births (LB)	Total births (TB= SB+LB)	All preg-nancies (TB+TOP)
					Number					
Vic	387	114	144	258	645	109	67	71	(138)	247
WA	169	76	59	135	304	88	4	19	(23)	111
SA	125	38	33	71	196	48	15	8	(23)	71
NSW[a]	230	142	141	283	513	67	53	84	(137)	204
Qld[b]	. .	29	122	151	. .	. .	52	44	96	. .
					Per cent					
Vic	60.0	17.7	22.3	(40.0)	100.0	44.1	27.1	28.7	(55.9)	100.0
WA	55.6	25.0	19.4	(44.4)	100.0	79.3	3.6	17.1	(20.7)	100.0
SA	63.8	19.4	16.8	(36.2)	100.0	67.6	21.1	11.3	(32.4)	100.0
NSW[a]	44.8	27.7	27.5	(55.2)	100.0	32.8	26.0	41.2	(67.2)	100.0
Qld[b]	. .	19.2	80.8	100.0	. .	. .	54.2	45.8	100.0	. .
					Prevalence[c]					
Vic	7.6	2.3	2.8	(5.1)	12.7	5.1	3.1	3.3	(6.5)	11.5
WA	8.3	3.7	2.9	(6.6)	14.9	9.8	0.4	2.1	(2.6)	12.4
SA	8.7	2.6	2.3	(4.9)	13.6	8.2	2.6	1.4	(3.9)	12.1
NSW[a]	3.3	2.0	2.0	(4.1)	7.4	2.3	1.9	2.9	(4.8)	7.2
Qld[b]	. .	0.7	3.0	3.8	. .	. .	2.9	2.5	5.4	. .

. . Indicates the value is not applicable

(a) In NSW, during 2006–2008, two babies born alive before 20 weeks gestation and weighing less than 400 grams have been included with TOP.

(b) There has been a change in criteria for data supplied by Queensland for NTD-affected births. For the years 1998–2005 all NTD-affected births that were at least 20 weeks gestation and 400 grams have been supplied. For the years 2006–2008 data has been supplied for all NTD-affected births with at least 20 weeks gestation or 400 grams birthweight.

(c) Prevalence is calculated by dividing the number of NTD by the number of total births in the population (Table A7.1). When applied to NTD from all pregnancies this is the overall prevalence. When applied to NTD from total births this is the birth prevalence. Prevalence is reported per 10,000 total births.

Notes

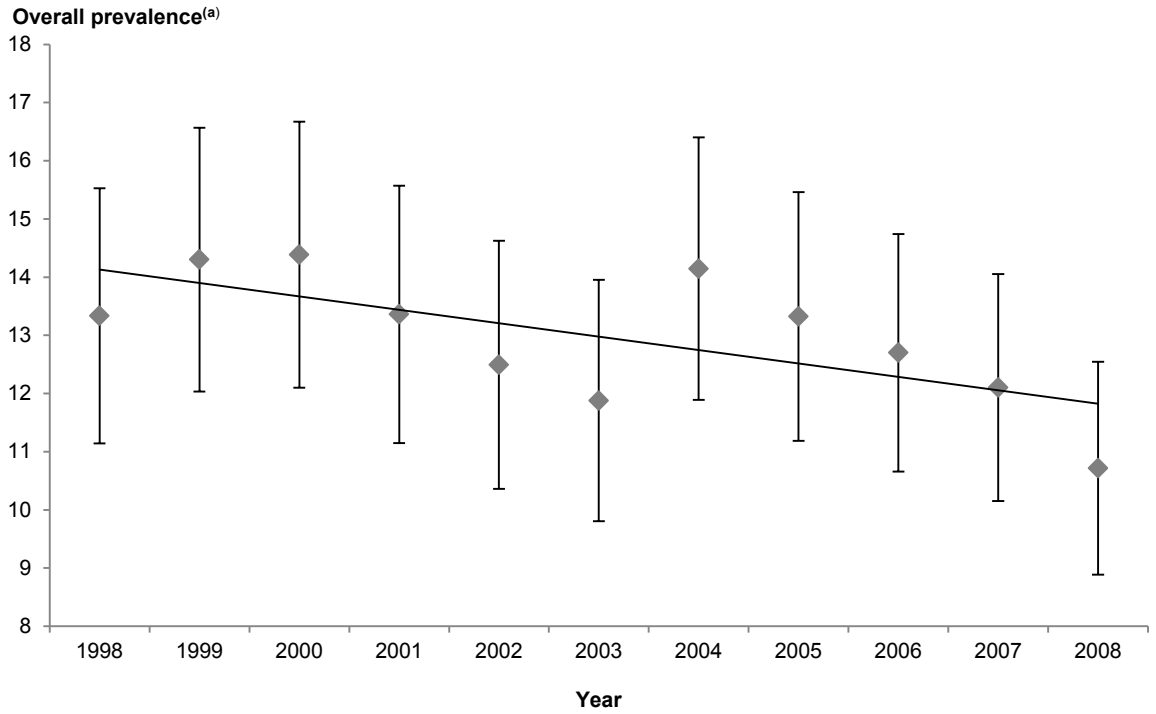
1. Termination of pregnancy (TOP) is defined as a termination of pregnancy for congenital anomaly before 20 weeks gestational age and weighing less than 400 grams.
2. Stillbirth (SB) is defined as a baby born with no sign of life of at least 20 weeks gestational age or 400 grams birthweight and includes terminations of pregnancy.
3. Live birth (LB) is a baby born of at least 20 weeks gestational age or 400 grams birthweight showing signs of life.
4. Percentage and prevalence estimates for births combine estimates in live births and stillbirths. Percentage and prevalence estimates for births and terminations of pregnancy include live births, stillbirths and terminations of pregnancy.
5. Number, per cent and prevalence of NTDs among births in Vic, WA and SA do not contribute independently to all pregnancies and are therefore shown in brackets.

3.2 Overall prevalence of neural tube defects

Overall prevalence of NTDs is estimated from jurisdictions that receive data both from births and from pregnancies that end in termination of pregnancy for congenital anomaly before 20 weeks gestational age. The overall prevalence of NTDs was calculated using data collated from the three states considered to have complete ascertainment of NTDs among pregnancies ending either as a birth or as a termination of pregnancy: Victoria, South Australia and Western Australia. For selected factors, an attenuated overall prevalence is reported including data from New South Wales.

Trends in overall prevalence of neural tube defect

Figure 3.1 shows overall prevalence of NTDs calculated for each calendar year together with the 95% confidence intervals (95%CI). In 1998 the overall prevalence of NTDs was 13.3 (95%CI: 11.1–15.5) per 10,000 total births. The lowest overall prevalence was recorded in 2008 when there were 10.7 (95%CI: 8.9–12.5) NTDs per 10,000 total births in 2008.



(a) Per 10,000 births.

Notes

1. Annual overall prevalence is indicated by the diamond marker.
2. Error bars indicate the 95% confidence intervals for each annual prevalence estimate. The line of best fit shown has the formula: y=14.2–0.23x.

Sources: ACAMS, NPDC and Table A6.1.

Figure 3.1: Overall prevalence of NTDs among pregnancies, Vic, WA and SA, 1998–2008

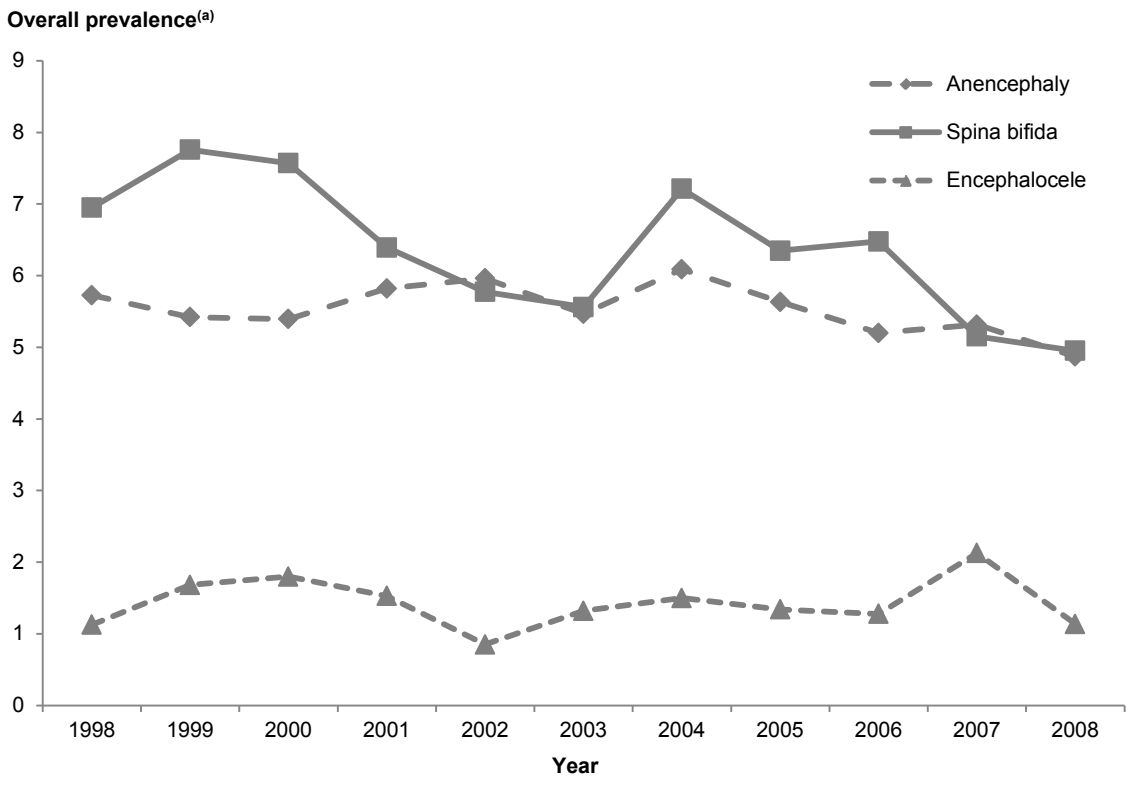
There have been fluctuations in the overall prevalence of NTDs over the period 1998–2008 (Figure 3.1). More precise estimates of overall prevalence of NTDs can be obtained by aggregating estimates over 3 years. From 2006 to 2008 there were 11.8 (95%CI: 10.7–12.9) NTDs per 10,000 births, compared with 14.0 (95%CI: 12.7–15.3) between 1998 and 2000. This

represents a decline in overall prevalence of NTDs across the whole period. Between 2006 and 2008 there were 2.2 (95%CI for difference 2.0–2.4) fewer NTDs per 10,000 total births than there were from 1998 to 2000.

Alternatively, this decline can be estimated by measuring the slope of the regression line shown in Figure 1, which represents the line that best fits the annual measures of overall prevalence of NTDs over the 11-year period. An average annual fall in overall prevalence of 0.2 per 10,000 was observed between 1998 and 2008 with a positive test for linear trend.

Trends in overall prevalence of spina bifida, anencephaly and encephalocele

Spina bifida and anencephaly have overall prevalence of a similar magnitude and are more common than encephalocele. Between 1998 and 2008, the overall prevalence of spina bifida was 6.3 (95%CI: 5.9–6.8) per 10,000 total births and non-significantly higher than the overall prevalence of anencephaly, which was 5.5 (95%CI: 5.1–5.9) per 10,000 total births. Encephalocele had the lowest overall prevalence of all NTDs from 1998 to 2008 of 1.4 (95%CI: 1.2–1.6) per 10,000 total births. The annual overall prevalence of anencephaly, spina bifida and encephalocele between 1998 and 2008 is presented in Figure 3.2.



(a) Per 10,000 births.

Sources: ACAMS, NPDC and Table A6.2.

Figure 3.2: Overall prevalence of NTDs, Vic, WA and SA, by year and type of condition

The net change in these conditions over the 11 years showed a more pronounced fall for spina bifida than for anencephaly or encephalocele. This is evident in the difference between the overall prevalence in the first 3 years between 1998 and 2000 and the last 3 years from

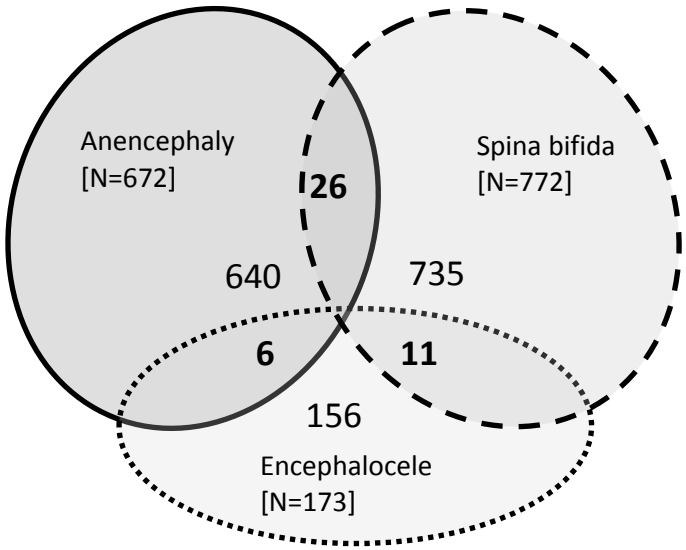
2006 to 2008. The overall prevalence of spina bifida showed a statistically significant decline from 7.4 (95%CI: 6.5–8.4) per 10,000 total births between 1998 and 2000 to 5.5 (95%CI: 4.8–6.3) between 2006 and 2008. There was a statistically non-significant reduction in the overall prevalence of anencephaly from 5.5 (95%CI: 4.7–6.3) per 10,000 total births from 1998 to 2000 to 5.1(95%CI: 4.4–5.9), while the overall prevalence of encephalocele was unchanged at 1.5 per 10,000 total births.

These changes can be summarised as average annual change in overall prevalence, which is obtained by applying a line of best fit to the annual estimates. The average annual reduction in overall prevalence of spina bifida was 0.20 per 10,000 total births, while for anencephaly the overall prevalence fell on average each year by 0.05 per 10,000 total births over this 11-year period. The overall prevalence of encephalocele rose on average annually by 0.01 per 10,000 total births.

The reduction in NTDs between 1998 and 2008 can be attributed largely to the reduction in overall prevalence of spina bifida.

Presence of more than one neural tube defect

In the 11 years from 1998 to 2008, there were 43 babies with more than one type of NTD. Among the 672 babies with anencephaly, 26 also had spina bifida and 6 encephalocele. There were 11 babies affected with both spina bifida and encephalocele. The number of babies with one or more reported NTDs is shown in Figure 3.3. There were no babies with all three types of NTD. Proportionally fewer babies with anencephaly (4.8%) or spina bifida (4.6%) had a second type of NTD than babies with encephalocele (10%).



Source: ACAMS

Figure 3.3: Babies with one or more NTDs, Vic, WA and SA, 1998–2008

The total number of neural tube defect conditions reported exceeds the number of babies affected by NTD. Where consistency between the prevalence of NTDs and the sum of the prevalence of types of NTD is required (that is, reporting is by baby and not by NTD condition) the hierarchical classification described above is applied yielding 672 cases classified as anencephaly, 746 as spina bifida (including 735 with spina bifida alone and 11 with spina bifida and encephalocele) and 156 (encephalocele only) as encephalocele.

Mothers and their babies affected by neural tube defects

This section considers trends in overall prevalence of NTDs by different characteristics of the mother or the baby. To accommodate the relatively small number of NTDs and follow the overall prevalence over time, a 9-year period was chosen so that results for the 1,279 affected babies could be aggregated into three triennial periods: 2000 to 2002, 2003 to 2005 and 2006 to 2008.

Preliminary examination of the data confirmed there were no pregnancies with more than one affected baby. Thus the estimates of overall prevalence and percentage of NTD apply equally to women who were pregnant and their babies.

Unless otherwise stated, the results presented in each table for each type of NTD include all reported cases. The final column, however, counts each woman or baby once. As with the figures above, the sum of the individual anomalies is greater than the number of affected women or babies. Where triennial results for NTD conditions are sparse the results for the 9-year period are presented.

Multiple pregnancy

The risk of having an NTD-affected pregnancy was increased for women with a multi-fetal pregnancy. In 9 years, from 2000 to 2008, the overall prevalence of NTDs was higher among multiple births with 22.8 per 10,000 multiple births compared with 12.8 per 10,000 singleton births (Table 3.2). This difference is statistically significant. The overall prevalence of NTDs among singleton pregnancies did not change substantially across the three triennial periods.

Table 3.2: Overall prevalence of neural tube defect according to pregnancy plurality, Vic, WA and SA, 2000–2008

	2000–2008			Any NTD			
	Anen-cephaly	Spina bifida	Enceph-alocele	2000 –2002	2003 –2005	2006 –2008	Total
				Number			
Singleton	527	603	139	409	413	418	1,240
Multiple	26	12	4	15	12	11	38
Not stated	—	—	1	—	1	—	2
				Overall prevalence[(a)(b)]			
Singleton	5.4	6.2	1.4	13.4	13.2	11.9	12.8
95% CI	5.0–5.9	5.7–6.7	1.2–1.7	12.1–14.7	11.9–14.5	10.8–13.1	12.1–13.5
Multiple	15.6	7.2	2.4	29.0	21.3	18.7	22.8
95% CI	9.6–21.6	3.1–11.3	0.0–4.7	14.3–43.6	9.3–33.4	7.7–29.8	15.5–30.0

(a) Per 10,000 total births.

(b) Denominator data can be found in Table A7.1.

Sources: ACAMS and NPDC.

In contrast, the overall prevalence of NTDs among multiple pregnancies showed a steady, but statistically non-significant, decline from 26.7 per 10,000 total births in 2000 to 2002 to 18.7 per 10,000 total births in 2006 to 2008.

The overall prevalence of anencephaly among multiple births was threefold higher than among singleton births. Examination of the trends over time for individual conditions are not presented in Table 3.2, but showed no change in the overall prevalence among singleton births. However, among multiple births there were statistically non-significant declines in the overall prevalence of anencephaly from 23.4 per 10,000 multiple births in 2000–2002 to 11.8 per 10,000 multiple births in 2006 to 2008 and spina bifida from 9.7 to 5.1 per 10,000 multiple births.

Neural tube defects as an isolated anomaly or one of multiple anomalies

In Victoria, South Australia and Western Australia between 2000 and 2008 there were 1,008 (79%) babies in whom NTD was assessed as an isolated anomaly. Among the babies with isolated NTD were 157 in whom additional reported anomalies were considered a consequence of, and not additional to, the NTDs and 7 in whom two types of NTD were present. Table 3.3 presents the number of affected babies, the percentage and overall prevalence of NTD-affected babies by classification as isolated NTD or multiple congenital anomalies that included at least one NTD together with one or more anomalies considered to be unrelated to the NTD.

Table 3.3: Overall prevalence of neural tube defect as an isolated anomaly or as one of multiple anomalies[(a)], Vic, WA and SA, over three triennia

	2000–2008			Any NTD			
	Anen-cephaly	Spina Bifida	Enceph-alocele	2000 –2002	2003 –2005	2006 –2008	Total
				Number			
Isolated anomaly	475	451	96	332	331	345	1,008
Multiple anomalies:[(a)]	75	149	43	85	91	74	250
Chromosomal	(16)	(41)	(4)	(17)	(20)	(21)	(58)
Other	(59)	(108)	(39)	(68)	(71)	(53)	(192)
Not determined	3	15	5	7	4	10	21
				Per cent			
Isolated anomaly	85.9	73.3	66.7	78.3	77.7	80.4	78.8
Multiple anomalies[(a)]:	13.6	24.2	29.9	20.0	21.4	17.2	19.5
Chromosomal	(2.9)	(6.7)	(2.8)	(4.0)	(4.7)	(4.9)	(4.5)
Other	(10.7)	(17.6)	(27.1)	(16.0)	(16.7)	(12.4)	(15.0)
Not determined	0.5	2.4	3.5	1.7	0.9	2.3	1.6
				Overall prevalence[(b)(c)]			
Isolated anomaly	4.7	4.5	1.0	10.5	10.2	9.5	10.0
Multiple anomalies[(a)]:	0.7	1.5	0.4	2.7	2.8	2.0	2.5
Chromosomal	(0.2)	(0.4)	(0.0)	(0.5)	(0.6)	(0.6)	(0.6)
Other	(0.6)	(1.1)	(0.4)	(2.2)	(2.2)	(1.5)	(1.9)

(a) Multiple anomalies indicate that in addition to the NTD there are one or more anomalies that are unrelated to the NTD. These are reported separately as chromosomal anomalies and other anomalies. Results for these categories appear in brackets and do not contribute independently to column totals.

(b) Per 10,000 total births.

(c) Denominator data can be found in Table A7.3.

Sources: ACAMS and NPDC.

One in five babies with NTD was assessed as having one or more additional anomalies considered to be unrelated to the NTDs. Over a quarter of these unrelated anomalies were chromosomal anomalies. The most common unrelated anomaly was a congenital musculoskeletal abdominal wall anomaly (ICD-10-AM 756.7). The full list of anomalies and their assignment as NTD-associated, unrelated to NTDs or as undetermined (could not be assessed without further information) is available in Appendix 5.

Anencephaly was the NTD most likely to be reported as an isolated anomaly. From 2000 to 2008, 85.9% of all babies with anencephaly had no additional unrelated anomaly reported, compared with 73.3% of babies with spina bifida and 66.7% of babies with encephalocele. There was no discernible trend in the percentage of babies with isolated NTD over the three triennia for babies with spina bifida or encephalocele. The percentage of babies with isolated anencephaly increased slightly with each triennium.

Maternal age

The overall prevalence of anencephaly, spina bifida and all NTDs among pregnancies was higher at the extremes of reproductive age, that is, among women aged less than 20 years and the women aged 35 years or more, than among women in the centre of the age distribution. Across the whole period, from 2000 to 2008, the highest overall prevalence of NTDs of 15.5 (95%CI: 11.6–19.5) per 10,000 total births was seen among women aged less than 20 years. The overall prevalence of NTDs declined steadily as a woman's age increased, reaching the lowest value among women aged 30–34 years. The overall prevalence of NTD-affected pregnancies among women aged 35 years rose again slightly to 13.9 (95%CI:12.3–15.5) per 10,000 total births (Table 3.4).

Table 3.4: Overall prevalence of neural tube defect by woman's age in years, Vic, WA and SA, 2000–2008

	2000–2008			Any NTD			
Age (years)	Anencephaly	Spina bifida	Encephalocele	2000–2002	2003–2005	2006–2008	Total
				Number			
Less than 20	27	32	5	17	18	25	60
20–24	79	97	27	71	62	62	195
25–29	161	169	29	121	119	112	352
30–34	174	158	48	127	124	124	375
35 and over	107	156	34	84	101	103	288
Not known	5	3	1	4	2	3	9
				Overall prevalence[(a)(b)]			
Less than 20	7.0	8.3	1.3	13.0	14.4	19.2	15.5
20–24	6.0	7.3	2.0	16.5	14.8	13.1	14.7
25–29	6.0	6.3	1.1	13.2	14.0	12.1	13.1
30–34	5.1	4.7	1.4	12.0	10.9	10.4	11.1
35 and over	5.2	7.5	1.6	14.7	15.3	12.2	13.9

(a) Per 10,000 total births.

(b) Denominator data can be found in Table A7.4.

Sources: ACAMS and NPDC.

Remoteness of area of usual residence

The remoteness of area of usual residence was available for 1,206 women that had NTD-affected pregnancy between 2000 and 2008. The overall prevalence of NTDs rose, as the area of residence was located further away from a major city. Across the whole period the overall prevalence of NTDs among women who lived in a major city was 11.7 (95%CI 10.9–12.5) per 10,000 total births, 13.4 (95%CI: 12.0–14.9) per 10,000 total births among women living in regional areas and 15.6 (95%CI: 11.1–20.2) per 10,000 total births among women in remote areas (Table 3.5). Table 3.5 shows the number of affected babies and the overall prevalence of NTDs according to the area of usual residence in the three triennia: 2000 to 2002, 2003 to 2005 and 2006 to 2008.

Within each of the three triennia there is a similar incremental increase in the overall prevalence of NTDs as the area of usual residence becomes more remote. Trends over time must be interpreted with caution in view of the relatively large number of records in the earlier years that did not include a postcode for usual residence.

Table 3.5: Overall prevalence of neural tube defect according to remoteness of area of usual residence, Vic, WA and SA, 2000–2008

	2000–2008			Any NTD			
Remoteness classification	Anen-cephaly	Spina bifida	Enceph-alocele	2000 –2002	2003 –2005	2006 –2008	Total
			Number				
Major cities	371	395	94	253	278	305	836
Regional	134	157	40	99	120	106	325
Remote	14	32	2	15	15	15	45
Not stated	34	31	8	57	13	3	73
			Overall prevalence[(a)(b)]				
Major cities	5.2	5.5	1.3	11.4	12.0	11.6	11.7
Regional	5.5	6.5	1.7	12.6	15.2	12.6	13.4
Remote	4.9	11.1	0.7	15.2	16.3	15.5	15.6

(a) Per 10,000 total births.

(b) Denominator data can be found in Table A7.5.

Sources: ACAMS and NPDC.

Survival of babies with neural tube defects

Table 3.6 presents the number and percentage of babies who died in utero before 20 weeks or after 20 weeks and as neonates, that is, within 28 days of birth. In Victoria, South Australia and Western Australia, the three states considered to have complete ascertainment between 2000 and 2008, only 182 (14%) of the 1,279 babies with NTDs were still alive 28 or more days after birth. The percentage of deaths with each NTD condition and for each of the three triennia shows that no babies survived the neonatal period with anencephaly. A quarter of all known babies with spina bifida or encephalocele survived beyond 28 days.

Table 3.6: NTD mortality by hierarchical classification of condition and over time, Vic, WA and SA, 2000–2008

	Fetal death[a]					
	Before 20 weeks	From 20 weeks	Neonatal death	All fetal and neonatal deaths	Neonatal survivor	Total
			Number			
Anencephaly	443	79	31	553	..	553
Spina bifida	246	157	46	449	149	598
Encephalocoele	56	35	4	95	33	128
Any NTD						
2000–2002	257	82	24	363	61	424
2003–2005	243	103	21	367	59	426
2006–2008	245	86	36	367	62	429
			Per cent			
Anencephaly	80	14.3	5.6	100.0	..	100.0
Spina bifida	41.1	26.3	7.7	75.1	..	100.0
Encephalocoele	43.8	27.3	3.1	74.2	..	100.0
Any NTD						
2000–2002	60.6	19.3	5.7	85.6	..	..
2003–2005	57	24.2	4.9	86.2	..	..
2006–2008	57	20	8.4	85.5	..	..
			Mortality ratio[b]			
Anencephaly	4.4	0.8	0.3	5.5	..	..
Spina bifida	2.5	1.6	0.5	4.5	..	..
Encephalocoele	0.6	0.3	0.0	0.9	..	..
Any NTD						
2000–2002	8.1	2.6	0.8	11.5	..	..
2003–2005	7.5	3.2	0.6	11.3	..	..
2006–2008	6.8	2.4	1.0	10.1	..	..

.. Indicates the value is not applicable

(a) Denominator data can be found in Table A7.3.

Notes

1. Fetal deaths after 20 weeks include terminations of pregnancy. Fetal deaths before 20 weeks include two deaths of babies born alive before 20 weeks gestational age and weighing less than 400 grams.
2. Neonatal deaths in the first 28 days among babies born after at least 20 weeks gestational age or 400 grams birthweight.
3. Mortality ratio is calculated as the number of deaths in each category divided by the number of total births in the corresponding time period and expressed per 10,000 live and stillbirths. Mortality ratios for stillbirths and neonatal deaths equate to the fetal and neonatal components respectively of the NTD-specific perinatal mortality rate.

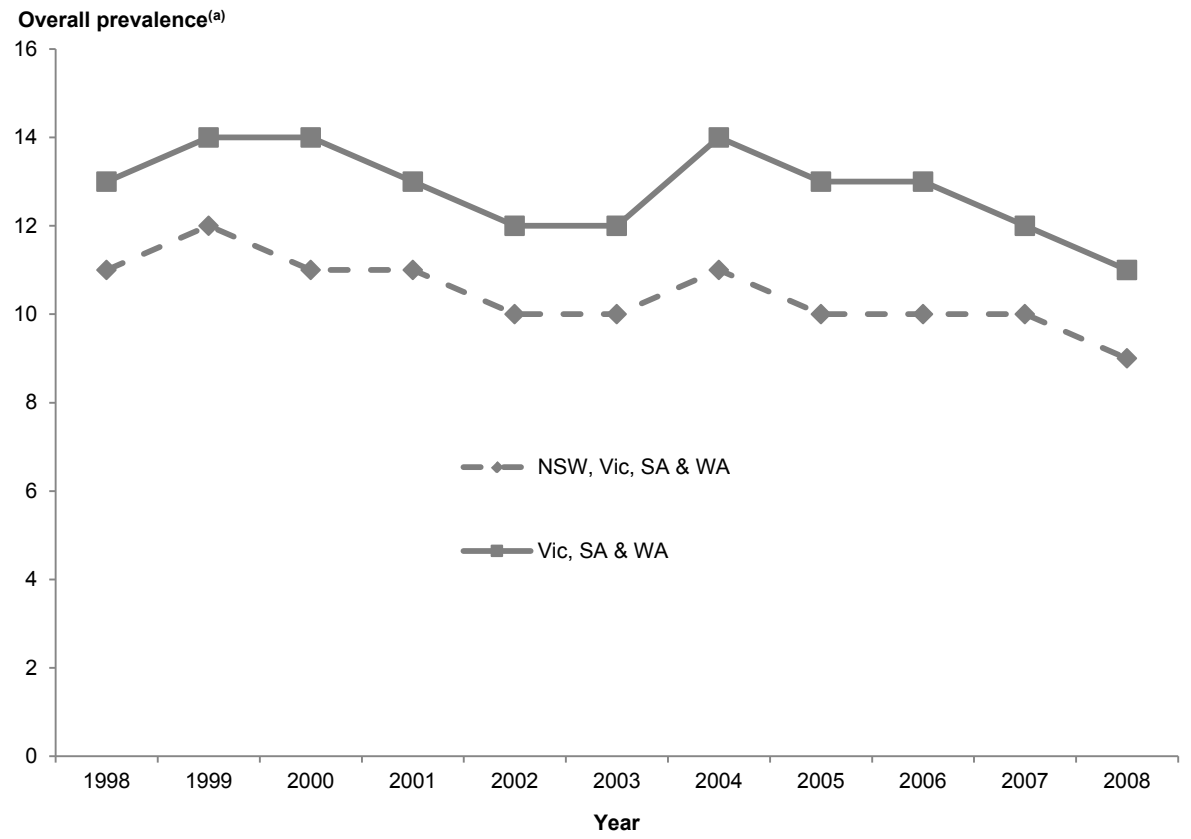
Sources: ACAMS and NPDC.

The percentage of babies with NTD who died was constant at 86% in the three triennia. The lower NTD mortality ratios evident over time do not reflect better survival of babies with NTD. Rather, these reflect the lower overall prevalence of NTDs over time.

These results do not provide much insight into the natural history and survival of babies with NTDs, as they are heavily influenced by the large proportion of pregnancies where NTDs were diagnosed prenatally and the pregnancy was terminated. Even among the babies born alive, the rates of neonatal survival may have been influenced by the decision to continue a pregnancy if a less severe form of the condition was diagnosed prenatally. The mortality ratios are not substantially different across the three triennia for anencephaly, spina bifida, encephalocele or any NTD.

3.3 Attenuated overall prevalence of neural tube defects

The overall prevalence of NTDs is lower when NSW data are included with data from Victoria, Western Australia and South Australia. The difference is shown in Figure 3.4, which compares the annual overall prevalence for the four states with the overall prevalence of NTDs for the three states. As the most populous state in Australia, it is advantageous to include NSW when considering certain population characteristics, even though the overall prevalence is attenuated.



(a) Per 10,000 births.

Sources: ACAMS, NPDC and Table A6.3.

Figure 3.4: Estimated overall prevalence of NTD, 1998–2008

Indigenous status

Indigenous status of the mother was available for just over 90% of all NTD-affected births and terminations of pregnancy in New South Wales and Western Australia. Indigenous status of the mother is not collected for pregnancies terminated for congenital anomaly in South Australia and is not of sufficient data quality from Victoria. Estimates of the overall prevalence of NTDs among women of Aboriginal and Torres Strait Islander origin are presented in Table 3.7 and are based on data from New South Wales and Western Australia.

The overall prevalence of NTDs in each of the three triennia and in all years among women of Aboriginal and/or Torres Strait Islander origin was twice that for women of non-Indigenous origin. From 2000 to 2008 this difference was statistically significant ($p<0.05$). The 86 records with missing information about the Indigenous status of the mother were not used to calculate these estimates. If some or any of these records relate to women of Aboriginal or Torres Strait Islander origin the overall prevalence estimated for this group would be even higher. If all of the records missing Indigenous status relate to women of non-Indigenous origin, the overall prevalence among non-Indigenous women would be little changed.

Table 3.7: Indigenous status of women that had an NTD-affected pregnancy, NSW and WA

	Any NTD			
	2000 –2002	2003 –2005	2006 –2008	Total
		Number		
Indigenous	15	22	23	60
Non-Indigenous	259	233	263	755
Not stated	27	30	29	86
		Overall prevalence[(a)]		
Indigenous	13.5	18.8	16.6	16.3
95% CI	6.7–20.3	10.9–26.6	9.8–23.4	12.2–20.5
Non-Indigenous	8.0	7.1	7.3	7.5
95% CI	7.0–9.0	6.2–8.0	6.4–8.2	6.9–8.0

(a) Per 10,000 total births.

(b) Denominator data can be found in Table A7.6.

Sources: ACAMS and NPDC.

Index of relative disadvantage

The SEIFA index of relative disadvantage was used to assign women to four groups according to the index of relative disadvantage calculated for their area of usual residence from information provided by households at each census. The overall prevalence of NTDs among pregnancies that ended in each of the census years and 1 year before and after was calculated for each quartile stratum and presented in Table 3.8.

During 2000 to 2002 the overall prevalence of NTD-affected pregnancies increased with the disadvantage level of the area of usual residence. For the years 2000 to 2002, women living in the least disadvantaged areas were observed to have the lowest overall prevalence of NTDs of 7.8 (95%CI: 6.5–9.2) per 10,000 total births. The overall prevalence of NTDs increased with

the level of disadvantage of the area of residence. Women living in the most disadvantaged areas were observed to have the highest overall prevalence observed in the period with 11.3 (95%CI: 9.6–12.9) NTDs per 10,000 total births.

Between 2005 and 2007, there was little difference in the overall prevalence of NTDs among women living in areas with the least disadvantage: 9.0 (95%CI: 7.6–10.3) per 10,000 total births and those living in areas with the most disadvantage 10.5 (95%CI: 8.8–12.3) per 10,000 total births.

Table 3.8: Overall prevalence of neural tube defect according to the area of residence level of disadvantage[(a)], NSW, Vic, WA and SA, in 2000–2002 and 2005–2007

	Number	Overall prevalence[(b,c)]	95% CI
2000–2002			
1st quartile—most disadvantaged	179	11.3	9.6–12.9
2nd quartile	122	9.0	7.4–10.6
3rd quartile	118	10.2	8.4–12.0
4th quartile—least disadvantaged	128	7.8	6.5–9.2
2005–2007			
1st quartile—most disadvantaged	141	10.5	8.8–12.3
2nd quartile	161	10.7	9.1–12.4
3rd quartile	156	10.3	8.7–11.9
4th quartile—least disadvantaged	174	9.0	7.6–10.3

(a) ABS Socioeconomic Index for Areas: Index of relative disadvantage.

(b) Per 10,000 total births.

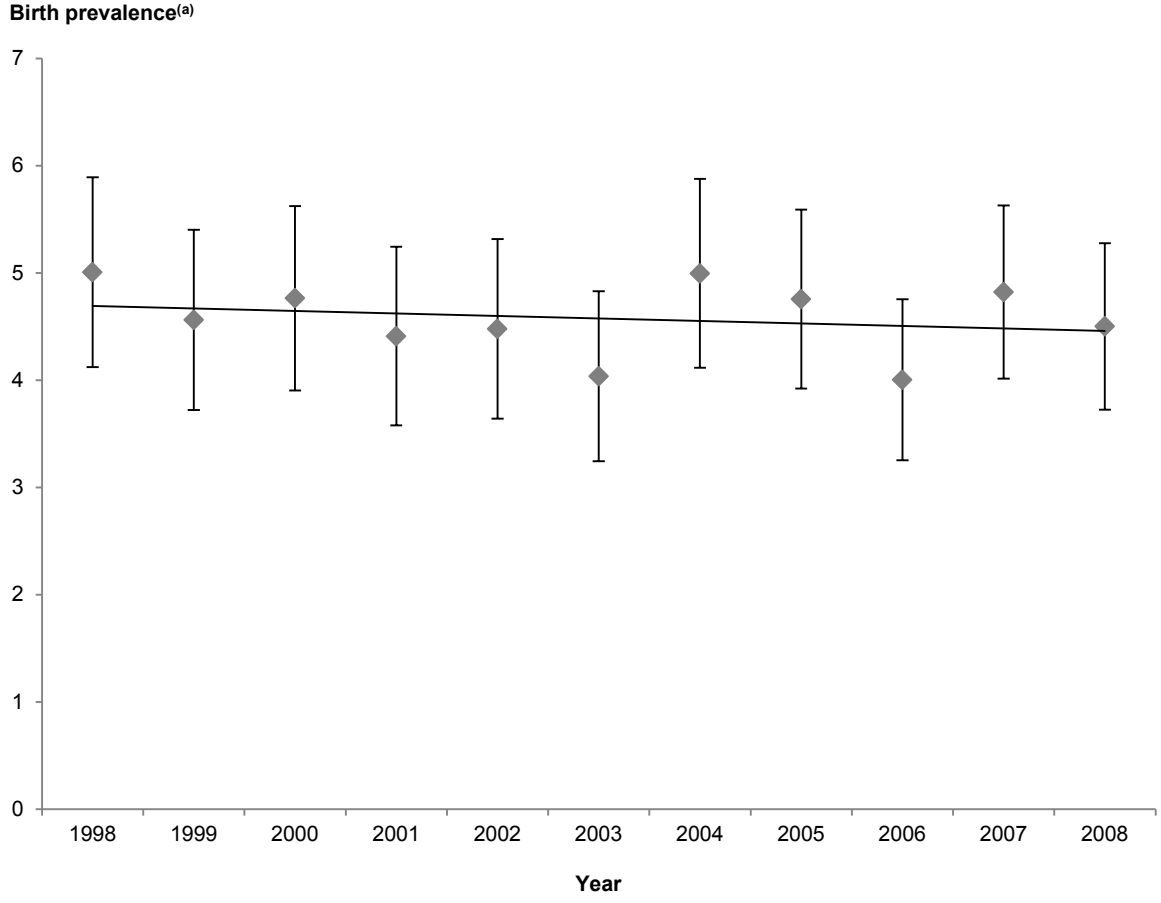
(c) Denominator data can be found in Table A7.8.

Sources: ACAMS and NPDC.

3.4 Birth prevalence of neural tube defects

Trends in birth prevalence

The birth prevalence of NTDs in the years 1998 to 2008 is shown in Figure 3.5 together with 95% confidence intervals around the annual estimates. The birth prevalence of NTDs at the end of the period in 2008 of 4.5 per 10,000 total births was not significantly different to that in 1998 when the birth prevalence was estimated to be 5.0 per 10,000 total births.



(a) Per 10,000 births.

Note: For the years 1998–2005 Queensland provided data for NTD-affected births that were at least 20 weeks gestation and 400 grams birthweight. Results for 2006–2008, exclude the additional NTD births obtained applying the criteria 20 weeks gestation or 400 grams birthweight so that annual birth prevalence is comparable over time.

Sources: ACAMS. NPDC and Table A6.4.

Figure 3.5: Birth prevalence of NTD-affected pregnancies, NSW, Vic, Qld, WA and SA 1998–2008

The birth prevalence of NTDs is determined both by the overall prevalence of NTDs and the extent to which NTD-affected pregnancies are terminated before 20 weeks gestational age. The latter is determined by the effectiveness of antenatal screening programs to detect fetal anomalies and the decisions made by those with a diagnosis of NTD about whether to continue with the pregnancy.

Birth prevalence in different states

The combined result for all states masks different trends in the annual birth prevalence of NTDs in different jurisdictions. Figure 3.6 shows the number of babies born each year with NTDs in each of the states that contributed data included in this report. In Victoria and New South Wales the birth prevalence over the most recent years has been increasing. The smaller states of Western Australia and South Australia have fewer births with NTDs and more fluctuation in the birth prevalence. The overall trend in the birth prevalence in these two jurisdictions over the 11 years is downwards.

Birth prevalence of neural tube defects among mothers and babies with different characteristics

Maternal characteristics

These data about characteristics of births with NTD update those in Abeywardana & Sullivan (2008b). Table 3.9 presents the number and birth prevalence of NTDs in each of the three triennia, 2000 to 2002, 2003 to 2005 and 2006 to 2008 among births with different maternal characteristics: age at birth, Indigenous status and maternal country of birth.

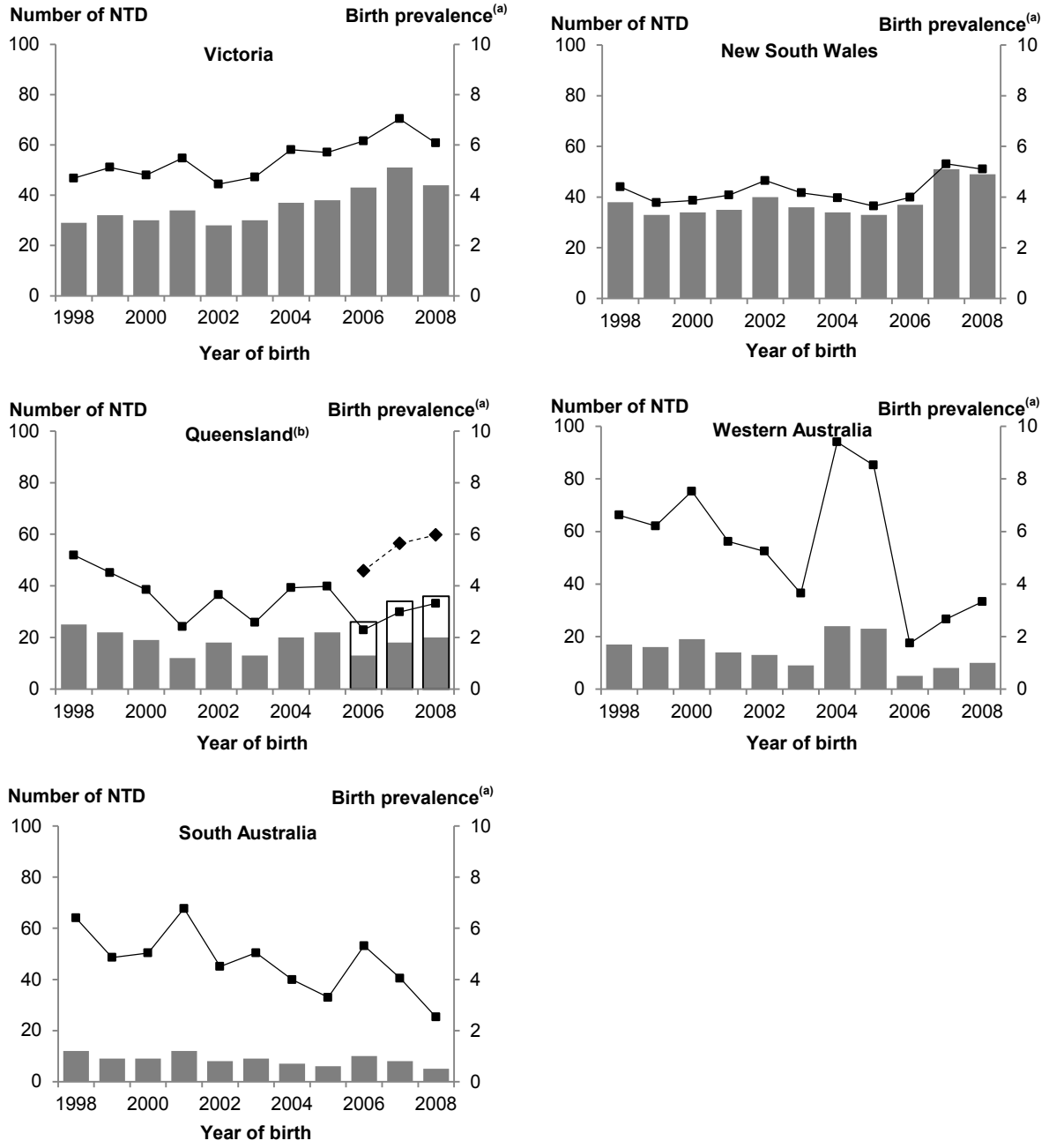
Women aged less than 25 years have consistently higher birth prevalence of NTDs in all triennia than women who are older. Across the three triennia the birth prevalence of NTDs increased among women aged less than 20 years and women aged 25–30 years at the time of birth. In contrast among women 30 years or older NTD birth prevalence decreased over time.

The birth prevalence of NTDs among women of Aboriginal and/or Torres Strait Islander origin in each of the three triennia was more than twice that found among non-Indigenous women. Fluctuation in the birth prevalence of NTDs would be expected as a result of the relatively small size of the Aboriginal and Torres Strait Islander population. Births in Victoria were omitted from this part of Table 3.9 because of known under-reporting of Indigenous status. The higher birth prevalence of NTDs among women of Aboriginal and Torres Strait Islander origin is evident in all triennia and among each NTD subtype.

Country of birth was available for almost 90% of NTD-affected pregnancies. The prevalence of NTDs among births to women born outside Australia was slightly higher than for women born in Australia. The prevalence for women born in Australia has decreased over the three triennia from 5.0 per 10,000 total births between 2000 and 2002 to 4.6 per 10,000 total births between 2006 and 2008. The prevalence of NTDs among women born outside Australia rose over the time period from 3.4 per 10,000 total births between 2000 and 2002 to 5.3 per 10,000 total births between 2006 and 2008.

Baby characteristics

Table 3.9 details characteristics of babies born with NTDs. Over the 9-year period, from 2000 to 2008, just over half (51%) of all births with NTDs occurred before 31 weeks gestational age. Terminations of pregnancy that were carried out after 20 weeks gestational age or more are included in this group as they are within the scope of the definition of a birth.



(a) Per 10,000 births.

(b) For the years 1998–2005 Queensland provided data for NTD-affected births that were at least 20 weeks gestation and 400 grams birthweight. The dashed line and the faded columns represent the prevalence and number of NTD-affected births when applying the original definitions. Results for 2006–2008, represented by the solid columns and line include the additional NTD births obtained applying the criteria 20 weeks gestation or 400 grams birthweight which are directly comparable to results in other jurisdictions.

Note: The axis scales, representing the number of NTD and the birth prevalence, are the same on each graph so the differences between states can be compared. The faded and filled columns represent the number of NTD and the solid and dashed lines represent birth prevalence.

Sources: ACAMS, NPDC and Table A6.5.

Figure 3.6: Number of NTD-affected births and birth prevalence by state, 1998–2008

Table 3.9: Neural tube defects by maternal characteristics, NSW, Vic, Qld, WA and SA, in three triennia: 2000–2002, 2002–2005 and 2006–2008

	2000–2008			Any NTD			
	Anen-cephaly	Spina bifida	Enceph-alocele	2000 –2002	2003 –2005	2006 –2008	Total
			Number				
Maternal age in years							
Less than 20	16	47	7	17	18	30	65
20–24	63	137	34	74	69	82	225
25–29	69	190	27	84	93	103	280
30–34	60	190	43	91	99	99	289
35 and over	34	139	26	56	61	78	195
Not known	4	22	4	3	1	25	29
Indigenous status[(a)]							
Indigenous	16	49	8	22	21	27	70
Not Indigenous	155	438	78	205	211	244	660
Not stated	5	14	—	6	4	8	18
Country of birth							
Australia	143	455	82	212	170	282	664
Not Australia	80	195	44	103	96	110	309
Not known	23	75	15	10	75	25	110
			Birth prevalence[(b)(c)]				
Maternal age in years							
Less than 20	1.6	4.7	0.7	5.0	5.6	9.0	6.5
20–24	1.9	4.1	1.0	6.7	6.4	6.9	6.7
25–29	1.1	3.0	0.4	3.9	4.6	4.7	4.4
30–34	0.8	2.5	0.6	3.9	3.9	3.7	3.8
35 and over	0.7	3.0	0.6	4.4	4.1	4.2	4.2
Indigenous status[(a)]							
Indigenous	2.4	7.2	1.2	10.6	9.6	10.7	10.3
Not Indigenous	0.9	2.7	0.5	3.9	4.0	4.1	4.0
Country of birth							
Australia	0.9	3.0	0.5	5.0	3.5	4.5	4.4
Not Australia	1.0	2.5	0.6	3.4	3.7	5.3	4.0

(a) Data from Victoria were not included because Indigenous status is not reliably reported.

(b) Per 10,000 births.

(c) Denominator data can be found in Table A7.9.

Sources: ACAMS and NPDC.

Birthweight was available for almost 97% of all NTD-affected births. The prevalence at birth of NTD-affected babies born weighing less than 1,500 grams rose by 27% since 2000–2002 with a birth prevalence of 181.2 per 10,000 total births between 2006 and 2008. The birth prevalence of babies born with a birthweight of 1,500–2,499 grams and 2,500 grams or more

remained at a similar level over the three triennia. These findings are likely to be linked with the changes in gestational age.

Table 3.10: Neural tube defects by baby characteristics, NSW, Vic, Qld, WA and SA, 2000–2008

	2000–2008			Any NTD			
	Anen-cephaly	Spina bifida	Enceph-alocele	2000 –2002	2003 –2005	2006 –2008	Total
				Number			
Gestational age at birth (completed weeks)							
20–31	157	355	62	152	167	232	551
32–36	42	63	14	40	33	43	116
37+	45	307	65	132	141	141	414
Not known	2	—	—	1	—	1	2
Birthweight							
<1,500	168	344	60	154	167	227	548
1,500–2,499g	40	60	14	34	41	36	111
≥2,500g	29	299	62	126	128	135	389
Unknown	9	22	5	11	5	19	35
Sex							
Male	115	365	70	165	171	206	542
Female	126	351	69	156	164	206	526
Not stated[(a)]	5	9	2	4	6	5	15
				Birth prevalence[(b)(c)]			
Gestational age at birth (completed weeks)							
20–31	41.4	93.6	16.4	129.7	135.5	167.3	145.3
32–36	2.9	4.4	1.0	9.0	7.0	8.1	8.0
37+	0.2	1.5	0.3	2.0	2.1	1.9	2.0
Birthweight							
<1,500	48.7	99.7	17.4	143.2	148.7	181.2	158.8
1,500–2,499g	3.3	5.0	1.2	8.9	10.4	8.4	9.2
≥2,500g	0.1	1.4	0.3	1.9	1.8	1.8	1.8
Sex							
Male	1.0	3.1	0.6	4.4	4.5	4.8	4.6
Female	1.1	3.1	0.6	4.4	4.5	5.1	4.7

(a) Sex not stated includes a small number of babies where sex was not determined at birth.

(b) Per 10,000 births. This has implications for monitoring mandatory folic acid fortification of bread flour.

(c) Denominator data can be found in Table A7.10.

Sources: ACAMS and NPDC.

The birth prevalence of female babies affected by any NTD is slightly higher than male babies for the three triennia. The rates have increased over this period from 4.4 males per 10,000 total births and 4.4 female per 10,000 total births between 2000 and 2002 compared with 4.9 male per 10,000 total births and 5.1 female per 10,000 total births between 2006 and

2008. The rates for the individual NTDs are very similar for each triennium with a consistently higher rate among females.

4 Key findings

The goal of mandatory fortification of bread flour with folic acid in Australia is the primary prevention of NTDs. Avoiding the occurrence of the mal-development of the central nervous system in the first weeks of embryonic life will lead to a decrease in the overall prevalence of NTD-affected pregnancies. This will directly benefit a small number of families who will have a healthy baby rather than a pregnancy or a baby affected by NTDs.

The benefit to the wider society stems from the savings brought about by a reduction in the number of babies with serious disability needing lifelong care (Dalziel et al. 2009; Tanya et al. 2008; Gross et al. 2008). A key measure of the impact of mandatory flour fortification with folic acid in Australia will be the extent of the reduction in the overall prevalence of NTDs in the post-fortification period.

Overall prevalence of neural tube defects

This report's findings provide a baseline from which to monitor the effect of mandatory flour fortification on the prevalence of NTDs. Additionally the results demonstrate three key aspects of the overall prevalence of NTDs that have implications for monitoring the effects of flour fortification:

- **A continued downward trend in the overall prevalence of NTDs in the 3 years since 2008.**

  Over the last 11 years there has been an average annual reduction of 0.22 NTDs per 10,000 total births. Factors that underlie this continuing declining overall prevalence of NTDs include, but are not restricted to, the use of folate supplements by women planning pregnancy, and the continuation of programs that directly or indirectly improve periconceptional nutrition (Bower & Stanley 2004) and changes in the socio-demographic structure of the maternity population (Laws, Li & Sullivan 2010).

  As a consequence any decrease in the overall prevalence of NTDs demonstrated in future years cannot be attributed wholly to the effects of mandatory flour fortification. Future assessment of the impact of mandatory folic acid fortification of bread flour should consider applying a correction factor to allow for the continuing decline in the overall prevalence of NTDs from other health promotion and NTD prevention programs.

- **Differences in trends in the overall prevalence of anencephaly, spina bifida and encephalocele.**

  Over the 11-year period there has been a fall in the overall prevalence of spina bifida, but no appreciable change in the overall prevalence of anencephaly or encephalocele. These three conditions have different patterns of association with other factors, particularly multiple pregnancy and the presence of other, unrelated anomalies. The changes in overall prevalence of these conditions after mandatory folic acid fortification in both Canada and the United States were not uniform. A more marked reduction in the overall prevalence of spina bifida than in either anencephaly or encephalocele was seen in both countries (Table 1.1). This has been attributed to the higher baseline for spina bifida (De Wals et al. 2007).

  When changes in NTD prevalence following mandatory fortification of bread flour with folic acid are monitored, it is important to consider the constituent conditions, particularly anencephaly and spina bifida, as well as all NTDs.

- **Unequal distribution of NTD overall prevalence across the population.**

  There was higher prevalence of NTD-affected pregnancies evident among women at both extremes of maternal age, but more so among younger women; among women living in areas of relative disadvantage; women living in remote areas; and among women of Aboriginal and Torres Strait Islander origin. These findings are consistent with the reported overall prevalence of NTDs in previous years (Abeywardana & Sullivan 2008b; Abeywardana et al. 2010; Bower et al. 2010; Gibson et al. 2009; Riley & Halliday 2008). These groups have already been identified as women who benefit least from current efforts to increase periconceptional folates through educational programs, the use of supplements or voluntary fortification of food (Abeywardana et al. 2010).

  The criteria for evaluation of mandatory flour fortification in Australia could, in addition to looking for a decline in overall prevalence of NTD, include provision for demonstrating less variation in the overall prevalence of NTDs among women of different maternal age, residential remoteness, residential area disadvantage and Indigenous origin. In the United States there has been a marked decrease in prevalence of NTDs in non-Hispanic black and non-Hispanic white women following mandatory flour fortification (Cordero et al. 2010). In Australia, it may not be possible to monitor the changes among Aboriginal and Torres Strait Islander women and those of non-Indigenous origin in the first phase of monitoring because of the relatively small size of Indigenous populations, but it would be feasible at a later stage by comparing prevalence over a longer time period.

Birth prevalence of neural tube defects

A secondary but nonetheless important outcome expected as a result of mandatory flour fortification will be the reduction in birth prevalence of NTDs. Changes in the birth prevalence are important to the final analysis as babies born with NTDs, in particular those who survive, have the greatest impact on society as a whole. Although NTDs are a rare set of conditions that affect very few families, these conditions may seriously affect quality of life for the individuals and their families, often incurring very high costs in terms of health care and other services required to promote survival and provision of long-term support.

It is not possible to quantify the change in birth prevalence attributable to mandatory flour fortification in jurisdictions that are unable to count termination of pregnancy. This is because birth prevalence depends on: the overall prevalence of NTDs in the population; the diagnostic effectiveness and uptake of programs for antenatal detection of NTDs; together with the individual choices women make about continuing a pregnancy that is found to be affected by NTD before 20 weeks gestation.

The two most relevant findings in this report:

- Birth prevalence of NTDs has remained relatively unchanged since 1998. Birth prevalence has remained constant over time and is of a similar order across jurisdictions. Western Australia stood out from 2006 to 2008 as having a lower birth prevalence that offset the higher prevalence among pregnancies terminated before 20 weeks gestation.
- Rising birth prevalence of NTDs is evident in some states. The higher birth prevalence of NTDs seen in the later years covered by this report is seen in the context of falling overall prevalence in Victoria and no evidence of change in the overall prevalence in NSW. This reflects the impact of individual decisions about whether or not to continue the pregnancy and the timing of pregnancy termination.

Methodological issues

The relative rarity of NTD and the small population size in the three jurisdictions that provided comprehensive data for this study in Australia pose particular challenges for measuring change. Just over 40% of births in Australia take place in Victoria, Western Australia and South Australia (Laws & Sullivan 2009). Monitoring changes in the overall prevalence of NTDs in these states will provide the most robust estimates of the overall prevalence of NTD. With qualifications, the additional cases from NSW can be used to monitor the effects particularly among women of Aboriginal and Torres Strait Islander origin.

A further consideration when measuring changes in the prevalence of conditions over relatively short time frames is the fact that NTD-affected pregnancies can vary substantially in duration. In short-term studies this can introduce truncation bias. Pregnancies that end in a termination before 20 weeks have half the duration of a pregnancy that reaches term. The critical exposure that the mandatory folic acid fortification of bread flour seeks to prevent occurs in the first weeks of pregnancy, near the time of conception. In this report, as in the previous report (Abeywardana & Sullivan 2008b) babies were grouped according to the year in which the birth occurred. In future reports assessing the effects of mandatory folic acid fortification of bread flour, babies should be grouped on the basis of the year in which the conception occurred to avoid truncation bias. This is an alternative to the method used in the American and Canadian studies (De Wals et al. 2007), in which cohorts were defined using the expected date of delivery.

Counts of NTD-affected pregnancies, as shown in this report, can be substantially altered by subtle changes in definition. This is evident with the data supplied by Queensland from 2006 to 2008. Data for 96 babies weighing 400 grams at birth or of 20 weeks or more gestation were supplied from 2006 to 2008. Data for only 54 babies would have been supplied if the rule requiring that both these conditions were met was again applied. This 83% increase in the number of NTD-affected births reported reflects the relatively large number of NTD-affected births at the lower gestational age range for births. NTD-affected babies may be small for gestational age and those affected by anencephaly are likely to weigh less because the skull and brain contribute a substantial proportion of the body mass. Although the definition of a birth is independent of the requirements for ascertaining NTD-affected babies, there is the potential for systematically omitting NTD-affected babies and in particular anencephalic babies born at or near 20 weeks gestational age if a minimum weight of 400 grams is also required. In this report for comparison of birth prevalence in Queensland over time the original definition was applied. Comparison of birth prevalence in Queensland with birth prevalence in other jurisdictions is only valid for births from 2006 to 2008.

Conclusion

This report provides the baseline prevalence of NTDs in Australia, before the national implementation of mandatory folic acid fortification of bread flour in September 2009. It also illustrates the current prevalence of NTDs and the component conditions in the context of trends in the rates of NTDs since 1998. It is expected that this will help evaluate the effect of mandatory folic acid fortification in the future.

Appendix 1: Definitions of neural tube defects

Neural tube defects

Neural tube defects comprise three distinct conditions: anencephaly, spina bifida and encephalocele.

Anencephaly

A congenital anomaly characterised by the total or partial absence of the cranial vault, the covering skin and the brain. Remaining brain tissue may be very much reduced in size. Craniorachischisis and iniencephaly are included, but acephaly, the absence of the head observed in amorphous acardiac twins, is excluded.

The following codes were used to select records with this condition:

ICD-9-BPA codes: 740.00–740.29 or ICD-10-AM codes: Q00.0–Q00.2.

Spina bifida

A congenital anomaly characterised by a failure in the closure of the spinal column and characterised by herniation or exposure of the spinal cord and/or meninges through the incompletely closed spine. This excludes spina bifida occulta and sacrococcygeal teratoma without dysraphism.

The following codes were used to select records with this condition:

ICD-9-BPA codes: 741.00–741.99 or ICD-10-AM codes: Q05.0–Q05.9.

Encephalocele

A congenital anomaly characterised by herniation of the brain and/or meninges through a defect in the skull.

The following codes were used to select records with this condition:

ICD-9-BPA codes: 742.00–742.09 or ICD-10-AM codes: Q01.0–Q01.2, Q01.8, Q01.9.

Appendix 2: State and territory congenital anomaly collections

New South Wales, Victoria, South Australia and Western Australia have congenital anomaly registers with multiple sources of ascertainment. In these states, data about NTD and congenital anomalies from perinatal data collections are supplemented by information from other hospitals, cytogenetic laboratories, perinatal death certificates, general practitioners and other doctors, autopsy reports and notifications of terminations of pregnancy. The governance arrangements, range of conditions included, ascertainment process and notification period vary between these four states.

The Victorian Birth Defects Register includes data on all congenital anomalies for all live births, stillbirths and terminations of pregnancy irrespective of age at diagnosis, with a period of detection up to 15 years of age. In Victoria, abortion legislation allows reporting of conditions for which pregnancy termination is carried out.

The Western Australia Register of Developmental Anomalies includes congenital anomalies diagnosed prenatally and in children up to 6 years of age. Validation studies in Victoria (Riley et al. 2004) and Western Australia (Bower et al. 2001) have shown these registers to be near complete.

The South Australian Birth Defects Register benefits from legislation mandating the notification of congenital anomalies in pregnancies irrespective of gestational age and in children up to 5 years of age. South Australian data is therefore considered almost complete. The New South Wales Register of Congenital Conditions relies on voluntary reporting of congenital anomalies detected during pregnancy or at birth, or diagnosed in infants up to 1 year of age.

In contrast, Queensland, Tasmania and the Australian Capital Territory rely on their perinatal data collections for information regarding NTD and other congenital anomalies that are present at birth and supplement this with information from hospital inpatient records.

The Northern Territory is drafting legislation that will include the collection of congenital anomaly data for public health purposes, but does not currently identify or collect these data.

Appendix 3: Data items requested from states and territories

Data Item	Requested for births	Requested for terminations of pregnancy
Admission date	✓	✓
Admitted patient care	✓	✓
Age at diagnosis	✓	✓
Age of mother at delivery	✓	
Age of woman at miscarriage or abortion		✓
Area of usual residence	✓	✓
Baby / fetus gestational age	✓	
Baby birth order	✓	
Baby date of birth	✓	
Baby's year of birth	✓	
Birth order of baby	✓	
Birth plurality	✓	
Birth status	✓	
Birthweight	✓	
Congenital anomaly code	✓	✓
Congenital anomaly number	✓	✓
Date of procedure	✓	✓
Date of miscarriage or abortion		✓
Establishment code	✓	✓
Fetal reporting order		✓
Hospital where miscarriage was treated or where abortion was carried out		✓
Induced abortion flag	✓	✓
Mother's date of birth	✓	
Mother's country of birth	✓	
Mother's Indigenous status	✓	
Mother's postcode	✓	
Mother's state or territory of usual residence	✓	
Multiple pregnancy	✓	✓
Number of previous abortions	✓	✓
Number of previous ectopic pregnancies	✓	✓
Number of previous live births	✓	✓
Number of previous pregnancies	✓	✓
Number of previous spontaneous abortions	✓	✓
Number of previous stillbirths	✓	✓
Parity	✓	✓
Separation mode	✓	
Setting of birth	✓	
Sex of baby	✓	
Source of notification	✓	✓
State or territory of birth	✓	
State or territory of miscarriage or abortion		✓
State or territory supplier	✓	✓
Status of baby at 28 days of age	✓	
Time of diagnosis	✓	
Woman's accommodation status		✓
Woman's country of birth		✓
Woman's date of birth		✓
Woman's id		✓
Woman's Indigenous status		✓
Woman's postcode		✓
Woman's state or territory of residence		✓
Year of miscarriage or abortion		✓

Appendix 4: Data elements used in the report

Sex

Was reported as male, female, indeterminate or not stated for live births and stillbirths, but not for terminations before 20 weeks gestational age.

Gestational age

The estimated gestational age was reported in completed weeks since the first day of the last menstrual period. This may have been calculated from dates, from ultrasound findings or determined by clinical assessment.

Birthweight

Birthweight is the first weight of the live born or stillborn baby obtained after birth, or the weight of the neonate or infant on the date admitted if this is different from the date of birth.

Maternal age at the end of the pregnancy

Maternal age at the end of the pregnancy was provided by some states or calculated by subtracting the mother's date of birth from the birth date of her baby, or the date on which the pregnancy ended.

Indigenous status of women

Women who identified as Aboriginal but not Torres Strait Islander origin, Torres Strait Islander but not Aboriginal origin, or Aboriginal and Torres Strait Islander origin were reported as being of Indigenous origin in this report. The category reported as non-Indigenous was of neither Aboriginal nor Torres Strait Islander origin. Not all jurisdictions were able to provide data for these women.

Postcode of usual residence

The postcode of usual residence of the woman was provided.

Plurality

Plurality refers to the number of fetuses or babies from a single pregnancy. In this report, the plurality category is 'singleton' for single births, and 'multiple' for twins, triplets, quadruplets, quintuplets and other higher order multiple pregnancy.

Diagnosis

Diagnosis refers to conditions that have been identified to a fetus or baby.

Birth status

Birth status refers to the status of the baby after the birth event.

Appendix 5: Neural tube defect-related and neural tube defect-unrelated conditions

Note

Only those conditions that were listed for a baby or pregnancy terminated for congenital anomaly and affected by NTDs in the years 2000–2008 were assigned as related conditions, unrelated conditions or as possibly related conditions.

This should not be viewed as a definitive list of related and unrelated conditions.

Conditions have been secondarily grouped for ease of reporting.

Neural tube defect - unrelated conditions

ICD-9-BPM CODE (to 4 characters)	Description
216.5	benign skin neoplasm
228.0–228.1	haemangioma other than skin
238.0	neoplasm of uncertain behaviour
270.1	phenylketonuria
389.9	unspecified deafness
524.0	anomaly of jaw size
593.7	vesico-ureteric reflux
744.0–744.8	congenital anomalies of ear, face and neck
745.0–745.6	bulbus cordis anomalies and anomalies of cardiac septal closure
746.0–746.9	anomalies of pulmonary valve
747.0–747.6	other congenital anomalies of circulatory system
748.0–748.6	congenital anomalies of respiratory system
749.0–749.2	cleft palate and cleft lip
750.1–750.3	other congenital anomalies of upper alimentary tract
751.0–751.7	other congenital anomalies of digestive system
752.0–752.9	congenital anomalies of genital organs
753.0–753.8	congenital anomalies of urinary system renal agenesis
755.0–755.5	other congenital anomalies of limbs
756.4–756.7	other congenital musculoskeletal anomalies
757.3–757.5	congenital anomalies of integument
758.0–758.8	chromosomal anomalies
759.0–759.9	anomalies of adrenal gland spleen anomaly
762.8	amniotic band syndrome
771.1–771.2	infections specific to the perinatal period

Neural tube defect - related conditions

ICD-9-BPM CODE	Description
214.0	lipoma
348.0	cerebral cysts
742.1–742.4	microcephalus
744.5–744.9	congenital anomalies of ear, face and neck
754.0–754.7	certain congenital musculoskeletal deformities
755.4–755.9	other congenital anomalies of limbs
756.1–756.3	other congenital musculoskeletal anomalies
759.1–759.2	anomalies of adrenal gland
778.0	hydrops fetalis
191.69	neoplasm of brain
225.30	benign Neoplasm of brain
743.10–743.19	microphthalmos
743.43	coloboma of iris
743.52–743.59	congenital anomalies of posterior segment
748.51–748.58	agenesis, hypoplasia and dysplasia of lung
756.00–756.06	anomalies of skull and face bones
756.81	other absent muscle - Other specified anomalies of muscle, tendon, fascia and connective tissue
756.88	other specified anomalies - Other specified anomalies of muscle, tendon, fascia and connective tissue

Undetermined conditions

ICD-9-BPM CODE	Description
756.0–756.8	other congenital musculoskeletal anomalies of skull and face bones
742.88	other specified anomalies of nervous system
376.50	enophthalmos
743.00–743.09	anophthalmos
756.09	unspecified anomalies of skull and face bones

Appendix 6: Data used in figures

Table A6.1: Overall prevalence of NTDs among pregnancies, 1998–2008

	Overall prevalence[(a)]	95% CI
1998	13.3	11.1–15.5
1999	14.3	12.0–16.6
2000	14.4	12.1–16.7
2001	13.4	11.1–15.6
2002	12.5	10.4–14.6
2003	11.9	9.8–14.0
2004	14.1	11.9–16.4
2005	13.3	11.2–15.5
2006	12.7	10.7–14.7
2007	12.1	10.2–14.1
2008	10.7	8.9–12.5

(a) Per 10,000 total births.

Sources: ACAMS, NPDC and Table A7.8.

Table A6.2: Overall prevalence[(a)] of NTDs by year, by type of condition, 1998–2008

	1998	1999	2000	2001	2002	2003	2004	2005	2006	2007	2008
Anencephaly	5.7	5.4	5.4	5.8	6.0	5.5	6.1	5.6	5.2	5.3	4.9
Spina bifida	6.9	7.8	7.6	6.4	5.8	5.6	7.2	6.3	6.5	5.2	5.0
Encephalocele	1.1	1.7	1.8	1.5	0.9	1.3	1.5	1.3	1.3	2.1	1.1

(a) Per 10,000 total births.

Sources: ACAMS, NPDC and Table A7.8.

Table A6.3: Estimated overall prevalence[(a)] of NTD, 1992–2008

	NSW, Vic, WA and SA	Vic, WA and SA
1992	15.0	18.0
1993	14.4	17.1
1994	14.2	19.3
1995	15.5	21.3
1996	13.2	17.6
1997	13.3	19.3
1998	11.3	13.3
1999	11.8	14.3
2000	11.4	14.4
2001	10.7	13.4
2002	9.9	12.5
2003	9.7	11.9
2004	10.7	14.1
2005	10.1	13.3
2006	10.1	12.7
2007	10.1	12.1
2008	9.2	10.7

(a) Per 10,000 total births.

Sources: ACAMS, NPDC and Table A7.8.

Table A6.4: Birth prevalence of NTD-affected pregnancies, 1998–2008

	Birth prevalence[(a)]	95% CI
1998	5.0	4.1–5.9
1999	4.6	3.7–5.4
2000	4.8	3.9–5.6
2001	4.4	3.6–5.2
2002	4.5	3.6–5.3
2003	4.0	3.2–4.8
2004	5.0	4.7–5.9
2005	4.8	3.9–5.6
2006	4.0	3.3–4.8
2007	4.8	4.0–5.6
2008	4.5	3.7–5.3

(a) Per 10,000 total births.

Sources: ACAMS, NPDC and Table A7.8.

Table A6.5: Number of NTD- affected births and birth prevalence by state, 1998–2008

	1998	1999	2000	2001	2002	2003	2004	2005	2006	2007	2008
						Number of NTD-affected births					
NSW	38	33	34	35	40	36	34	33	37	51	49
Vic	29	32	30	34	28	30	37	38	43	51	45
Qld[(a)]	25	22	19	12	18	13	20	22	13	18	20
Qld	. .	. .	. .	. .	. .	. .	. .	. .	26	34	36
WA	17	16	19	14	13	9	24	23	5	8	10
SA	12	9	9	12	8	9	7	6	10	8	5
						Birth prevalence[(b)]					
NSW	4.4	3.8	3.9	4.1	4.7	4.2	4.0	3.6	4.0	5.3	5.1
Vic	4.7	5.1	4.8	5.5	4.4	4.7	5.8	5.7	6.2	7.0	6.2
Qld[(a)]	5.2	4.5	3.9	2.4	3.7	2.6	3.9	4.0	2.3	3.0	3.3
Qld	. .	. .	. .	. .	. .	. .	. .	. .	4.6	5.6	6.0
WA	6.6	6.2	7.5	5.6	5.2	3.6	9.4	8.5	1.7	2.7	3.3
SA	6.4	4.9	5.0	6.8	4.5	5.0	4.0	3.3	5.3	4.1	2.5

(a) Contains births that were at least 20 weeks gestation and 400 grams birthweight.

(b) Per 10,000 total births.

Sources: ACAMS, NPDC and Table A7.8.

Appendix 7: Denominator data

Table A7.1: Number of births, 1998–2008

	Number of births
1998–2005	
Victoria	506,532
Western Australia	203,592
South Australia	144,133
New South Wales	696,027
Queensland	401,667
2006–2008	
Victoria	214,874
Western Australia	89,412
South Australia	58,522
New South Wales	285,120
Queensland	178,352

Source: NPDC.

Table A7.2: Number of births by plurality, Vic, WA and SA, 2000–2008

	2000–2002	2003–2005	2006–2008	2000–2008
Singleton	305,659	313,310	350,962	969,931
Multiple	5,178	5,626	5,874	16,678

Source: NPDC.

Table A7.3: Number of all births, 2000–2008

	2000–2002	2003–2005	2006–2008	2000–2008
Vic, WA and SA	316,119	324,658	362,808	1,003,585
NSW, Qld , Vic, WA and SA	724,106	743,862	826,280	2,294,248

Source: NPDC.

Table A7.4: Number of women who gave birth by age group, Vic, WA and SA, 2000–2008

	2000–2002	2003–2005	2006–2008	2000–2008
<20	13,071	12,479	13,042	38,592
20–24	43,085	41,925	47,368	132,378
25–29	91,404	84,756	92,588	268,748
30–34	106,017	113,535	119,639	339,191
35+	57,246	66,215	84,176	207,637

Source: NPDC.

Table A7.5: Remoteness of the area of residence, women who gave birth, Vic, WA and SA, 2000–2008

	2000–2002	2003–2005	2006–2008	2000–2008
Major cities	222,186	230,871	262,726	715,783
Regional	78,735	78,780	84,136	241,651
Remote	9,872	9,185	9,699	28,756

Source: NPDC.

Table A7.6: Indigenous status of woman who gave birth, NSW and WA, 2000–2008

	2000–2002	2003–2005	2006–2008	2000–2008
Indigenous	11,135	11,729	13,836	36,700
Not Indigenous	323,457	327,943	359,747	1,011,147

Source: NPDC.

Table A7.7: Number of total births by SEIFA, NSW, Vic, WA and SA, 2000–2002 and 2005–2007

	2000–2002	2005–2007
SEIFA – NSW, Vic, WA, SA		
1st quartile—most disadvantaged	158,932	133,990
2nd quartile	135,562	149,812
3rd quartile	115,710	151,465
4th quartile—least disadvantaged	163,555	194,217

Source: NPDC.

Table A7.8: Number of births, NSW, Vic, Qld, WA and SA, 1998–2008

	1998	1999	2000	2001	2002	2003	2004	2005	2006	2007	2008
NSW	86,305	87,289	87,922	85,858	86,005	86,414	85,626	90,608	92,768	96,016	96,336
Vic	62,091	62,689	62,564	62,149	63,133	63,552	63,700	66,654	69,856	72,472	72,546
Qld	48,163	48,746	49,316	49,690	49,196	50,366	50,910	55,280	56,708	60,244	61,400
WA	25,677	25,771	25,229	24,939	24,784	24,681	25,528	26,983	28,664	30,074	30,674
SA	18,733	18,519	17,872	17,704	17,745	17,844	17,521	18,195	18,802	19,751	19,969

Source: NPDC.

Table A7.9: Characteristics of women who gave birth, NSW, Vic, Qld, WA, and SA, 2000–2008

	2000–2002	2003–2005	2006–2008	2000–2008
Age group (years)				
<20	34,259	32,205	33,411	99,875
20–24	110,087	107,930	118,703	336,720
25–29	217,966	203,061	221,054	642,081
30–34	234,816	255,137	270,326	760,279
35+	128,541	147,474	185,614	461,629
Indigenous status[(a)]				
Indigenous	20,681	21,972	25,184	67,837
Not Indigenous	520,228	533,305	591,893	1,645,426
Country of birth				
Australian	419,999	483,375	620,164	1,523,538
Not Australian	301,997	259,030	206,036	767,063

(a) Data from Victoria were not included.

Source: NPDC.

Table A7.10: Characteristics of births, NSW, Vic, Qld, WA, and SA, 2000–2008

	2000–2002	2003–2005	2006–2008	2000–2008
Gestational age (weeks)				
20–31	11,720	12,328	13,865	37,913
32–36	44,466	46,954	53,361	144,781
37+	667,838	684,503	758,935	2,111,276
Birthweight (grams)				
<1,500	10,755	11,230	12,531	34,516
1500–2499	38,070	39,294	42,917	120,281
2500+	675,076	693,053	770,434	2,138,563
Sex				
Male	371,455	382,252	424,796	1,178,503
Female	352,390	361,306	401,180	1,114,876

Source: NPDC.

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