Congenital anomalies in Australia 1998–2001

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Congenital anomalies in Australia 1998–2001

Samanthi Abeywardana
Muhsin Karim
Narelle Grayson
Elizabeth A Sullivan

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Any enquiries about or comments on this publication should be directed to:

AIHW National Perinatal Statistics Unit

Sydney Children's Hospital

Level 2 McNevin Dickson Building

Randwick Hospitals Campus

Randwick NSW 2031

AUSTRALIA

Phone: (02) 9382 1014

Email: npsu@unsw.edu.au

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1 Introduction

Congenital anomalies are a significant health concern in Australia, and remain a major reason for hospitalisation in infancy and childhood. They often result in disabilities and handicaps and, in some cases, death. An estimated 5.0% of all Australian births and terminations of pregnancy have a major congenital anomaly (Stanley 1995). Birth anomalies account for about 13.0% of the disease burden for children aged 0–14 years (AIHW 1999). Congenital anomalies are also a leading cause of infant mortality in Australia, with 25.0% of infant deaths in 2000 caused by birth anomalies (Al-Yaman et al. 2002). Perinatal deaths due to congenital anomalies affected 15.6 per 10,000 births in 1996, the lowest level since 1973 (Hurst et al. 1999).

A congenital anomaly is a structural, functional, chromosomal or metabolic abnormality that is present at birth, even if not diagnosed until months or years later. Most congenital anomalies originate in the embryonic period (up to the 7th week of gestation) and the others in fetal life (8th week to term).

The National Perinatal Statistics Unit has collected data from all states and territories since 1981 and reported it in the Birth Defects Series until 1997. The data have been provided by the state and territory health authorities primarily from their birth defects registers and congenital anomalies and perinatal data collections. There is variability among the states and territories in the scope of their congenital anomalies data collections, the sources of notification of anomalies, the definitions and classifications used, the method of data collection and the available resources.

A review report published in 2004 highlighted the value of high quality national data and recommended developing a new system. A collection of good quality data will enable the surveillance of congenital anomalies within the population over time. The data collected over a number of years enable a large sample size to be achieved which is useful for the study of less prevalent anomalies.

The following work is being undertaken for the development of the new system:

Developing a minimum dataset for congenital anomalies

Developing clinical definitions for congenital anomalies

Mapping classifications used for coding

Reporting congenital anomalies up to one year of age from all states and territories

Reporting termination of pregnancies irrespective of gestational age from all states and territories

Meanwhile, the 1998–2001 congenital anomalies report compiles the available data from states and territories to update national information on selected congenital anomalies in 1998 to 2001. This report analyses 33 congenital anomalies in detail. It reports characteristics and outcomes of the births as well as demographic and pregnancy characteristics of their mothers. Information on terminations of pregnancy before 20 weeks of gestation for congenital anomalies is also presented for some jurisdictions. The data provided for this report are not comparable among the states and territories because of the differences in the data collections in each jurisdiction. This has been considered in the presentation of the data and information has been provided to assist with interpretation. The Northern Territory data are not included for this report as they are not resourced to provide the data to the expected standard. Therefore the Northern Territory population is not included in the denominator for calculations.

Aims of the Australian Congenital Anomalies Monitoring System

- Improve ascertainment and quality of data by providing a framework and setting to standardise definitions, classifications and collection methods among the states and territories.
- Provide high quality, nationally consistent data for use in policy development and planning, including identifying areas of need and funding requirements.
- Enable smaller states and territories to merge with larger data collections for more meaningful statistics (e.g. increases the sample size).
- Enable national compilation and reporting of conditions (e.g. genetic disorders) not currently compiled and reported at the national level.
- Enable the evaluation of national health promotion activities aimed at preventing congenital anomalies.
- Enable research at the national level, including assisting with planning and managing collaborative research.
- Facilitate Australia's participation in international reporting and research.
- Provide a context for the evaluation of reported clusters of birth anomalies.
- Assist in reducing the burden of disease within the Australian population.

2 Data sources and methods

Australian Congenital Anomalies Monitoring System

The data are based on notifications to birth defects registers in New South Wales, Victoria, Western Australia and South Australia, and on data collected on congenital anomalies in Queensland, Tasmania and the Australian Capital Territory. The Northern Territory was unable to provide unit record data on congenital anomalies. Therefore, no data for the Northern Territory are included in this report.

The state and territory health authorities undertake data processing, analysis and publication of reports. Each state and the Australian Capital Territory provided data in an electronic format to NPSU. Due to editing and subsequent updates of state and territory databases, the numbers in this report may differ from those published in reports by the states and territories. See Appendix B for a list of state and territory reports.

Births in 1998–2001 were reported to the National Perinatal Data Collection (NPDC) by the states and territories and many of the data elements requested for inclusion in the Australian Congenital Anomalies Monitoring System (ACAMS) are also included in the NPDC. Therefore, to avoid duplication of effort, the states and territories were asked to provide a key to match the births with congenital anomalies reported to the ACAMS to their birth record in the NPDC. Victoria and South Australia were able to provide this information. The data elements were extracted from the NPDC for the births for these two states.

Scope

Criteria for inclusion

Births (live births and stillbirths ≥20 weeks gestational age or ≥400g birthweight) and terminations of pregnancy (TOPs, <20 weeks gestational age) with a congenital anomaly, occurring in 1998–2001 are included in this report (Table 1.1). Terminations of pregnancy at ≥20 weeks gestational age or ≥400g birthweight with a congenital anomaly are recorded as births. Those terminations of pregnancy ≥20 weeks gestational age or ≥400g birthweight were recorded as still births in all states and territories. Only some states and territories can recognise stillbirths and terminations of pregnancy separately.

Periods of notification

The prevalence of some congenital anomalies is affected by the availability and use of prenatal screening programs and diagnostic testing services, and whether the results of prenatal diagnostic tests are notified to congenital anomaly collections. The period of detection varies considerably among the state and territory collections and ranges from prenatal diagnosis to detection up to 15 years of age. All states and territories except Queensland, Australian Capital Territory and Tasmania have a period of notification up to one year of age or beyond. Queensland and Tasmania data collections include only the anomalies detected during the birth episode prior to discharge from the hospital. Queensland notifies prenatally diagnosed anomalies only if they are apparent at birth.

The prevalence of some congenital anomalies is also affected by whether induced abortions following prenatal diagnosis of a congenital anomaly are undertaken, and whether these congenital anomalies are notified to congenital anomalies data collections. Currently, the extent of the notification is not clear for terminations of pregnancy with congenital anomalies. The states collecting data on terminations of pregnancy at <20 weeks gestation may have incomplete data.

For this report, the data for New South Wales include births and induced abortions (<20 weeks gestational age) with congenital anomalies notified up to one year of age. The data for Victoria, Western Australia and South Australia include births and terminations of pregnancy(<20 weeks gestational age) with congenital anomalies notified up to 15 years, 6 years and 5 years of age respectively. The data for the remaining states and territories (Queensland, Tasmania and the Australia Capital Territory) include births with congenital anomalies notified in the perinatal period.

Terminations of pregnancy at ≥20 weeks gestational age or ≥400g birthweight due to congenital anomalies although recorded as births, can be identified separately from births for all jurisdictions except New South Wales and Tasmania.

Northern Territory data are not included in this report.

Table 1.1: Notification of terminations of pregnancy for congenital anomalies, by state and territory, 1998–2001

Notifications		NSW	Vic	Qld	WA	SA	Tas	ACT	NT
Induced abortion < 20 weeks following prenatal diagnosis of a congenital anomaly	Yes	✓	✓		✓	✓			
	No			✓			✓	✓	✓
Induced abortion ≥ 20 weeks following prenatal diagnosis of a congenital anomaly	Yes	✓	✓	✓	✓	✓	✓		
	No							✓	✓

Sources of notification

Table 1.2: Notifications of congenital anomalies, by state and territory, 1998–2001

Source of notification	NSW	Vic	Qld	WA	SA	Tas	ACT	NT
Routine data collections								
Hospital morbidity data	✓	✓		✓	✓		✓	✓
Perinatal/midwives data	✓	✓	✓	✓	✓	✓	✓	✓
Death certificates/mortality data		✓	✓	✓	✓			
Primary healthcare staff								
Notification by hospitals/clinicians/lab	✓			✓	✓			
Referral hospital record reviews	✓				✓			
General practitioners		✓		✓				
Obstetrician		✓		✓	✓			
Early childhood centre staff		✓		✓				
Rural healthcare workers				✓				
Other		✓			✓			
Disability services staff				✓	✓			
Medical officers								
Paediatricians	✓	✓	✓	✓	✓			
Other	✓	✓	✓	✓	✓			
Screening and diagnosis services								
Prenatal diagnosis (clinician/lab)	✓	✓		✓				
Cytogenetic/pathology reports	✓	✓	✓	✓	✓			
Ultrasound reports		✓		✓				
Newborn/genetic screening/diagnosis reports	✓	✓		✓	✓			
Other (e.g. parent)	✓	✓		✓				
Autopsy reports	✓			✓	✓			
Induced abortion data		✓		✓	✓			
Unknown	✓	✓						

Selected conditions included for national reporting

Congenital anomalies

The congenital anomalies presented in this report were agreed by the National Congenital Anomalies Steering Committee (NCASC). The selection is based on the 35 'sentinel' conditions reported on by the International Clearinghouse for Birth Defects Surveillance and Research (ICBDSR). Some of the conditions reported by the ICBDSR are not included in this report or are presented differently. Undescended testis and prune belly sequence are not included because NCASC agreed that they are difficult to define. Unspecified abdominal wall defects are not presented.

The descriptions of the conditions are based on those developed by ICBDSR and by the Victorian Birth Defects Register.

Classification of congenital anomalies

The classification used to code congenital anomalies varied among the states and territories and among years for some jurisdictions. For New South Wales, Victoria, Queensland, Western Australia and South Australia, congenital anomalies were classified using the Royal College of Paediatrics and Child Health's (formerly the British Paediatric Association (BPA)) Classification of Diseases (BPA 1979) for 1998–2001. For Tasmania, ICD-9-BPA was used for most cases in 1998 to 2001 and the International Statistical Classification of Diseases and Related Health Problems, 10th Revision; Australian Modification (ICD-10-AM) (NCCH 1998) was used for a small number of cases. NPSU, in consultation with Tasmania determined the equivalent ICD-9-BPA codes for these congenital anomalies and updated the records with these codes.

For the Australian Capital Territory, congenital anomalies were coded using the International Classification of Diseases, 9th Revision, Clinical Modification (ICD-9-CM) (NCC 1996) and ICD-10-AM for all records each year. The ICD-9-CM codes were used for the conditions for which ICD-9-BPA and ICD-9-CM are equivalent at the 4-character level. The ICD-10-AM codes were used for arhinenecephaly/holoprosencephaly, microtia, epispadias, hypospadias diaphragmatic hernia, exomphalos and gastroschisis.

The International Classification of Diseases (ICD) was developed by the World Health Organization (WHO) to enable international comparability for mortality statistics. The clinical modification of the 9th Revision of ICD was developed in the USA and provided a better classification for morbidity statistics. The National Coding Centre (NCC) developed the Australian version of ICD-9-CM in 1995.

The ICD is hierarchical, with a small number of disease chapters, including one on congenital malformations, that are divided into a large number of more specific disease groupings (represented by three character codes) which in turn can mostly be divided into an even larger number of more specific disease categories, represented by four and five character codes.

The ICD-9-BPA is an adaptation of ICD-9. It is an expansion of ICD-9 where more detail is required for the classification to be useful for paediatrics. A fifth character has been added to the fourth character level of ICD-9 to provide greater precision and specificity where necessary. It is also a contraction of ICD-9 in that it omits codes for conditions not commonly encountered in paediatrics.

New South Wales, Victoria, Queensland, Western Australia and South Australia have extended ICD-9-BPA to further specify some congenital anomalies.

Measures reported

In Chapter 3, information is presented on babies with the selected congenital anomalies and on women who gave birth to babies with the selected congenital anomalies. The number of babies is marginally higher than the number of women who gave birth because of multiple births.

Reported rate

Information on live births and fetal deaths of ≥20 weeks gestation or ≥400g birthweight is available for all states and territories. Induced abortions of ≥20 weeks gestation or ≥400g birthweight are included as births. The measure based on live births provides information about the burden of disease and disability associated with the selected congenital anomaly. The measure based on live births and fetal deaths provides information about the rate of the selected congenital anomaly among all births. Information on termination of pregnancy at <20 weeks gestation is available for four states: New South Wales, Victoria, Western Australia and South Australia. The rates for Top <20 weeks are provided as estimated rates because the denominator includes only the births and reported fetal deaths.

The numerator and denominator values used to derive the rates for each congenital anomaly are described in Table 1.3 and Table 1.4.

Table 1.3: Numerators and denominators used for calculating birth rates from all states and territories

Numerator[(a)]	Denominator[(b)]
All births	
Number of live births and fetal deaths with the selected congenital anomaly	Number of live births and fetal deaths
Live births	
Number of live births with the selected congenital anomaly	Number of live births
Fetal deaths	
Number of fetal deaths with the selected congenital anomaly	Number of fetal deaths
Women who gave birth	
Number of women who gave birth to babies with the selected congenital anomaly	Number of women who gave birth to a liveborn or stillborn baby

(a) Reported to the Australian Congenital Anomalies Monitoring System.

(b) Reported to the National Perinatal Data Collection (see Appendix A).

Table 1.4: Numerators and denominators used for calculating birth and termination of pregnancy rates from states providing data on TOP <20 weeks

Numerator[(a)]	Denominator[(b)]
Births and termination of pregnancies	
Number of live births, fetal deaths and termination of pregnancies[(c)] with the selected congenital anomaly	Number of live births and fetal deaths
Women who gave birth or who had a termination of pregnancy	
Number of women who gave birth to a baby with the selected congenital anomaly or who had a termination of pregnancy[(c)] for the selected congenital anomaly	Number of women who gave birth to a liveborn or stillborn baby

(a) Reported to the Australian Congenital Anomalies Monitoring System.

(b) Reported to the National Perinatal Data Collection (see Appendix A).

(c) Terminations of pregnancy at <20 weeks gestation.

Data elements included for national reporting

Characteristics of babies and of their mothers are presented for each congenital anomaly in Chapter 3. For babies, sex, gestational age and birthweight are presented. For women who gave birth, age group, Indigenous status, Remoteness area of usual residence, country of birth, parity and plurality are presented.

The data elements are defined in or are based on data elements defined in the National Health Data Dictionary (NHDD).

Notes on data elements – baby characteristics

Sex

Data on the sex of each baby were reported as male, female, indeterminate, not stated.

Gestational age

The estimated gestational age of the baby in completed weeks. This may be calculated from the first day of the last menstrual period or be determined by clinical assessment. WHO identifies preterm as less than 37 completed weeks of gestation, term as 37 completed weeks to less than 42 completed weeks of gestation and post-term as 42 completed weeks or more of gestation. Preterm birth is associated with morbidity and mortality in newborn babies.

For this report, the duration of the mother's pregnancy rather than the gestational age of the baby was requested from the states and territories. Gestational age is reported for births, so for multiple births, the gestational age of the first born baby was used for subsequent babies.

Birthweight

The first weight of the liveborn 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.

The weight of infants is only recorded if they are admitted to hospital and if they weigh ≤9,000g and are aged <365 days. For perinatal collections the birthweight is provided for all liveborn and stillborn babies.

For this report, it was requested that the weight at birth be provided for all records. For some cases, where the congenital anomaly was notified after the perinatal period, the birthweight may not be available but for some states and territories may have been obtained by linking to their perinatal data collection.

Birthweight is an indicator of a baby's health status. Babies are defined as being of low birthweight if their weight at birth is less than 2,500g.

Notes on data elements – Maternal characteristics

Age

The AIHW National Perinatal Statistics Unit calculated the age of mothers at the birth by subtracting their date of birth from the date of birth of their baby. Maternal age is presented by five-year age groups. Some congenital anomalies are associated with mothers in younger or older age groups.

Indigenous status

The Indigenous status of the mother was provided by the states and territories.

In this report, Indigenous status categories included as Indigenous mothers who were Aboriginal but not Torres Strait Islander origin, Torres Strait Islander but not Aboriginal origin, or Aboriginal and Torres Strait Islander origin. The category reported as non-Indigenous was neither Aboriginal nor Torres Strait Islander origin.

Remoteness Area of usual residence

The NHDD specifies that data on usual residence should be provided as the state or territory and the Statistical Local Area (SLA) of usual residence. For this report, data on the area of usual residence of mothers were provided as state/territory and SLA for Victoria, South Australia and the Australian Capital Territory. For Victoria and South Australia, state/territory and SLA was only provided if the woman's usual residence was in Victoria or South Australia respectively. The postcode was provided for all states and territories. For this report, postcodes have been assigned to Remoteness Areas to enable reporting of women who gave birth by Remoteness Area of usual residence. This is based on the Australian Statistical Geographical Classification (ASGC) remoteness structure (ABS 2001).

Because of the small numbers in the data presented, the Remoteness Areas of usual residence categories in this report are presented under three headings; major cities, regional and remote. (Inner regional and Outer regional are reported as Regional; Remote and Very remote are reported as Remote)

Country of birth

Country of birth details were requested to be provided using the Australian Bureau of Statistic's (ABS) Australian Standard Classification of Countries for Social Statistics (ASCCSS) (ABS 1990). All states and territories except New South Wales and Queensland provided country of birth details using the ASCCSS. The New South Wales data for country of birth, which were provided using the Standard Australian Classification of Countries (SACC) (ABS 1998), were mapped by the National Perinatal Statistics Unit to ASCCSS. For Queensland, country of birth details were provided using 20 defined groups.

Parity

The number of previous pregnancies that resulted in live births or stillbirths. In this report, the category included as primiparous was the first pregnancy and the categories included as multiparous were coded as one to eight or more. This data element was not specified in the NHDD in 1998–2001. For 1998 to 2001 New South Wales did not provide data on parity of the mother.

Plurality

The number of babies resulting from a single pregnancy. In this report, the plurality category included as singleton was *Singleton* and the categories included as multiple were twins, triplets, quadruplets, quintuplets, sextuplets and other. This data element was not specified in the NHDD in 1998–2001.

Australian Paediatric Surveillance Unit

The Australian Paediatric Surveillance Unit (APSU) facilitates active surveillance of uncommon childhood diseases, complications of common diseases or adverse effects of treatment. Diseases are chosen for their public health significance and impact on health resources.

APSU undertakes active surveillance of some conditions on a monthly basis. Clinicians working in paediatrics and child health throughout Australia participate and are asked to complete a report card for all children newly diagnosed with any of the conditions that are listed on the card. Study investigators for particular conditions then send a brief questionnaire to the clinician requesting further de-identified information. Investigators are responsible for collation, analysis and publication of these data and report study findings annually to the APSU.

Individuals or organisations may apply to study a condition through the APSU and applications undergo a process of peer review and revision before being included on the monthly report card. To satisfy the criteria for study, the condition must:

- be sufficiently uncommon so that the system is not over-burdened
- usually result in referral to a paediatrician or related specialist
- provide information that satisfies the study aims and that is not available from other sources.

Conditions are usually studied for one to three years.

It was a recommendation of the review of the National Congenital Malformations and Birth Defects Data Collection (Birch et al. 2004) that data from the ACAMS should be supplemented by data from other sources, such as data collected by the APSU. In consultation with the APSU, it was agreed that data on Prader-Willi syndrome (genetic), fetal alcohol syndrome (preventable), CHARGE association, congenital arthrogryposis and congenital rubella/varicella (preventable) could be included in this report. Data on these conditions are presented in Chapter 4.

State and territory comparisons

There are differences among the states and territories in the scope of their data collections. In particular, terminations of pregnancy at <20 weeks gestation are not included for Queensland, Tasmania and the Australian Capital Territory. The prevalence of conditions for which termination of pregnancy following prenatal diagnosis is a treatment option may be underestimated in these jurisdictions. Therefore, data have not been presented by state and territory for separate conditions.

Conventions

Throughout the report, for totals, percentages may not add up to 100.0, and subtotals may not add up to the sum of the percentages for the categories. This is due to rounding.

Minor changes to data presentation, including where a jurisdiction has not provided a data item or data have not been published for other reasons, are detailed in the footnotes to the tables.

3 Selected congenital anomalies

Introduction

This chapter presents data on 33 selected congenital anomalies. As noted in Chapter 1, the selection of the congenital anomalies was based on those reported to the International Clearinghouse for Birth Defects Surveillance and Research (ICBDSR) and was agreed by the National Congenital Anomalies Steering Committee (NCASC).

Data are not presented by state and territory in this report because of the variation in the period of notification in the data collected from different jurisdictions. Information given on fetal deaths includes stillbirths and terminations at or after 20 weeks of pregnancy.

The total number of the anomalies reported here does not represent the total number of the babies who had congenital anomalies. Some babies had more than one anomaly and the same baby will be counted for separate anomalies. Therefore, the total number of conditions will exceed the total number of babies who had congenital anomalies. For the same reason, the induced abortions reported may not be related to the anomaly given.

Estimated rates presented are based on the data provided from New South Wales, Victoria, South Australia and Western Australia. These four states provided data on congenital anomalies detected at birth (live births, stillbirths and induced abortions at 20 or more weeks of pregnancy) as well as the terminations of pregnancy before 20 weeks of gestation due to congenital anomalies. However terminations of pregnancy at <20 weeks may not be complete except for South Australia where reporting of all terminations of pregnancy is mandatory.

The parity of the mother was not provided by New South Wales and data on parity are based on the data from all other states. Tasmania does not collect data on Indigenous status; hence the denominator data do not include Tasmania for this data element.

Baby's sex, gestational age and birthweights are presented only for births at or more than 20 weeks of pregnancy. Those births include live births, stillbirths and terminations of pregnancy at ≥20 weeks. The details for these data elements were not available for terminations of pregnancy at <20 weeks gestation.

Anencephaly

Definition: A congenital anomaly characterised by the total or partial absence of the cranial vault, the covering skin, and the brain missing or reduced to a small mass. Include infants with craniorachischisis, iniencephaly and other neural tube defects such as encephalocele or open spina bifida, when associated with anencephaly. Exclude acephaly, that is, absence of head observed in amorphous acardiac twins.

ICD-9-BPA codes: 740.00–740.29

Anencephaly is not compatible with life and is usually diagnosed and terminated before birth. Many cases are terminated in early pregnancy. There was a slight reduction in reported cases at birth from 1998 to 2001 for all births (Table 2.1.1). The data provided by four states that include termination of pregnancy before 20 weeks shows that 4.5 per 10,000 births were affected by this anomaly (Table 2.1.2). The rate of babies born with anencephaly was equal among both sexes. The mean gestational age at birth was 28 completed weeks. The mean birthweight was 1,120g and 84.4% were under 2,500g (Table 2.1.3).

The mean maternal age of women who gave birth to a baby with anencephaly was 28.4 years. Most of those women (82.6%) were in 20–35 years age group. Indigenous women and women who had multiple births had a higher rate of births with this anomaly. More women living in remote areas were affected by a pregnancy with an anencephaly than the women living in cities.

Reported rates of anencephaly

Table 2.1.1: Number and rate of anencephaly by outcome, gestation ≥20 weeks or birthweight ≥400g, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Live births	17	0.7	8	0.3	9	0.4	7	0.3	41	0.4
Fetal deaths	18	10.4	15	8.6	18	10.1	17	9.8	68	9.8
All births	35	1.4	23	0.9	27	1.1	24	1.0	109	1.1

(a) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths.

Table 2.1.2: Number and estimated rate of anencephaly by outcome, gestation ≥20 weeks or birthweight ≥400g and TOP <20 weeks, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Live births	9	0.5	5	0.3	7	0.4	5	0.	26	0.3
Fetal deaths	16	12.4	12	9.2	18	13.6	15	11.7	61	11.8
All births	25	1.3	17	0.9	25	1.3	20	1.0	87	1.1
Births and TOP[(c)]	99	5.1	82	4.2	84	4.3	84	4.4	349	4.5

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths. For births and terminations of pregnancy, the rate is per 10,000 live births and fetal deaths.

(c) Terminations of pregnancy <20 weeks gestation. TOP ≥ 20 weeks gestation or ≥ 400g birthweight are included as births.

Baby characteristics

Table 2.1.3: Births (gestation ≥20 weeks or birthweight ≥400g) with anencephaly by baby characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 births
Sex			
Male	54	49.5	1.0
Female	53	48.6	1.1
Indeterminate	1	0.9	. .
Not stated	1	0.9	. .
Gestational age (weeks)			
20–36	85	78.0	11.0
37–41	20	18.3	0.2
42 and over	3	2.8	1.6
Not stated	1	0.9	. .
Birthweight (grams)			
Less than 2,500	92	84.4	13.7
2,500–4,499	13	11.9	0.1
Not stated	4	3.7	. .

Maternal characteristics

Table 2.1.4: Number of women who gave birth to babies with anencephaly, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Women who gave birth	35	1.4	23	0.9	27	1.1	24	1.0	109	1.1

(a) The rate is per 10,000 women who gave birth.

Table 2.1.5: Number of women who gave birth to babies with anencephaly or who had a TOP where a fetus had anencephaly and the estimated rate, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Women who gave birth[(c)]	25	1.3	17	0.9	25	1.3	20	1.1	87	1.1
Women who gave birth or had a TOP[(d)]	98	5.2	81	4.2	84	4.4	84	4.5	347	4.6

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) The rate is per 10,000 women who gave birth.

(c) Includes women who had a termination of pregnancy at ≥ 20 weeks gestation or ≥ 400g birthweight.

(d) Women who had a termination of pregnancy at <20 weeks gestation.

Table 2.1.6: Births (gestation ≥ 20 weeks or birthweight ≥ 400g) with anencephaly by maternal characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 women
Maternal age group			
Less than 20	6	5.5	1.2
20–24	25	22.9	1.6
25–29	32	29.4	1.0
30–34	27	24.8	0.9
35–39	13	11.9	0.9
40 and over	5	4.6	1.9
Indigenous status[(1)]			
Indigenous	10	9.3	3.4
Non-Indigenous	94	87.9	1.0
Not stated	3	2.8	. .
Remoteness Area			
Major cities	74	67.9	1.2
Regional	27	24.8	1.0
Remote	5	4.6	2.2
Not stated	3	2.8	. .
Country of birth			
Australia	81	74.3	1.3
New Zealand	3	2.8	2.1
United Kingdom	2	1.8	0.6
Europe	1	0.9	0.4
Middle East and North Africa	5	4.6	2.7
Asia	10	9.2	1.4
Africa (excluding North Africa)	1	0.9	1.4
Other countries	1	0.9	1.1
Not stated	5	4.6	. .
Parity[(2)]			
Primiparous	22	31.0	0.8
Multiparous	47	66.2	1.2
Not stated	2	2.8	. .
Plurality			
Singleton	87	79.8	0.9
Multiple	22	20.2	13.9

(1) Excludes data from Tasmania

(2) Excludes data from New South Wales

Spina bifida

Definition: A family of congenital anomaly defects in the closure of the spinal column characterised by herniation or exposure of the spinal cord and/or meninges through an incompletely closed spine. Include meningocele, meningomyelocele, myelocele, myelomeningocele and rachischisis. Spina bifida is not counted when present with anencephaly. Exclude spina bifida occulta, sacrococcygeal teratoma without dysraphism.

ICD-9-BPA codes: 741.00–741.99

The rate of spina bifida at birth remained consistent, with an overall rate of 3.1 per 10,000 births (Table2.2.1). The data provided by four states that included termination of pregnancy before 20 weeks show that 6.1 per 10,000 births were affected by this anomaly (Table 2.2.2). There were more males than females affected with spina bifida. The mean gestational age at birth was 31 completed weeks. The mean birthweight for the babies with spina bifida was 2058g and half of them weighed below 2500g (Table 2.2.3). Of the affected cases 93% were singleton births.

The average maternal age was 27.6 years and young women <25 years of age had a higher rate of affected pregnancies. The rate of having an affected baby with this anomaly was more than three times higher for Indigenous women; 9.3 vs 2.9/10,000 women who gave birth. Women living in remote areas had a higher rate of spina bifida births than the women living in cities. More multiple births than singleton births were affected with this anomaly (Table 2.2.6).

Reported rates of Spina bifida

Table 2.2.1: Number and rate of spina bifida by outcome, gestation ≥20 weeks or birthweight ≥400g Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Live births	46	1.8	52	2.1	59	2.3	42	1.7	199	2.0
Fetal deaths	32	18.5	29	16.6	25	14.1	31	17.9	119	17.1
All births	80	3.2	81	3.2	84	3.3	73	2.9	318	3.1

(a) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths.

Table 2.2.2: Number and the estimated rate of spina bifida by outcome, gestation ≥20 weeks or birthweight ≥400g and TOP <20 weeks, available for four states[(a)] only,1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Live births	37	1.9	36	1.9	44	2.3	36	1.9	153	2.0
Fetal deaths	26	20.2	26	20.0	21	15.9	29	22.6	104	20.0
All births	65	3.4	62	3.2	65	3.4	65	3.4	257	3.3
Births and TOP[(c)]	114	5.9	133	6.8	119	6.1	103	5.4	469	6.1

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths. For births and terminations of pregnancy, the rate is per 10,000 live births and fetal deaths.

(c) Termination of pregnancy <20 weeks gestation. TOP ≥ 20 weeks gestation or ≥ 400g birthweight are included as births.

Baby characteristics

Table 2.2.3: Births (gestation ≥20 weeks or birthweight ≥400g) with spina bifida by baby characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 births
Sex			
Male	167	52.5	3.2
Female	147	46.2	3.0
Indeterminate	2	0.6	. .
Not stated	2	0.6	. .
Gestational age (weeks)			
20–36	168	52.8	21.7
37–41	146	45.9	1.6
42 and over	4	1.3	2.2
Birthweight (grams)			
Less than 2,500	157	49.4	23.4
2,500–4,499	145	45.6	1.6
4,500 and over	5	1.6	2.7
Not stated	11	3.5	. .

Maternal characteristics

Table 2.2.4: Number of women who gave birth to babies with spina bifida, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Women who gave birth	80	3.2	81	3.2	84	3.4	73	3.0	318	3.2

(a) The rate is per 10,000 women who gave birth.

Table 2.2.5: Number of women who gave birth to babies with spina bifida or who had a TOP where a fetus had spina bifida and the estimated rate, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Women who gave birth[(c)]	65	3.4	62	3.2	65	3.4	65	3.5	257	3.4
Women who gave birth or had a TOP[(d)]	114	6.0	133	7.0	119	6.2	103	5.5	469	6.2

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) The rate is per 10,000 women who gave birth.

(c) Includes women who had a termination of pregnancy at ≥ 20 weeks gestation or ≥ 400g birthweight.

(d) Women who had a termination of pregnancy at <20 weeks gestation.

Table 2.2.6: Births (gestation ≥ 20 weeks or birthweight ≥ 400g) with spina bifida by maternal characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 women
Maternal age group			
Less than 20	28	8.8	5.8
20–24	72	22.6	4.6
25–29	102	32.1	3.3
30–34	66	20.8	2.1
35–39	38	11.9	2.7
40 and over	8	2.5	3.1
Not stated	4	1.3	. .
Indigenous status[1]			
Indigenous	27	8.7	9.3
Non-Indigenous	274	88.1	2.9
Not stated	10	3.2	. .
Remoteness Area			
Major cities	189	59.4	3.1
Regional	97	30.5	3.7
Remote	19	6.0	8.4
Not stated	13	4.1	. .
Country of birth			
Australia	257	80.8	4.2
New Zealand	7	2.2	4.9
United Kingdom	11	3.5	3.4
Europe	6	1.9	2.7
Middle East and North Africa	9	2.8	4.9
Asia	9	2.8	1.3
North America	1	0.3	2.0
Other countries	1	0.3	1.1
Not stated	17	5.3	. .
Parity[2]			
Primiparous	78	35.6	3.0
Multiparous	134	61.2	3.5
Not stated	7	3.2	. .
Plurality			
Singleton	296	93.1	3.0
Multiple	22	6.9	13.9

(1) Excludes data from Tasmania

(2) Excludes data from New South Wales

Encephalocele

Definition: A congenital anomaly characterised by herniation of the brain and/or meninges through a defect in the skull. Encephalocele is not counted when present with spina bifida or anencephaly.

ICD-9-BPA codes: 742.00–742.09

The rate of encephalocele in births was 0.7 per 10,000 births from 1998 to 2001 (Table 2.3.1). The data from the states providing data on termination of pregnancy before 20 weeks shows a higher rate of affected births (Table 2.3.2).

Of the reported cases there were more females than males (Table 2.3.3) and mean gestational age at birth was 31 completed weeks. The mean birthweight for those babies was 1,991g and 59.1% had a birthweight less than 2,500 grams.

The number of women affected by a pregnancy with encephalocele during the four year period was 93 and the mean maternal age was 27.9 years. Most of those women were aged <35 years (88%). The rates of encephalocele births did not differ markedly between Indigenous and non-Indigenous women and also among women living in remote areas compared to the women living in cities. The rate of encephalocele births was higher for births among multiparous women and for multiple births.

Reported rates of Encephalocele

Table 2.3.1: Number and rate of encephalocele by outcome, gestation ≥20 weeks or birthweight ≥400g, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Live births	10	0.4	12	0.5	8	0.3	11	0.4	41	0.4
Fetal deaths	7	4.1	5	2.9	7	3.9	6	3.5	25	3.6
All births	17	0.7	17	0.7	15	0.6	17	0.7	66	0.7

(a) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths.

Table 2.3.2: Number and the estimated rate of encephalocele by outcome, gestation ≥20 weeks or birthweight ≥400g and TOP <20 weeks, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Live births	5	0.3	9	0.5	5	0.3	7	0.4	26	0.3
Fetal deaths	6	4.7	5	3.8	5	3.8	6	4.7	22	4.2
All births	11	0.6	14	0.7	10	0.5	13	0.7	48	0.6
Births and TOP[(c)]	19	1.0	25	1.3	29	1.5	21	1.1	94	1.2

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths. For births and terminations of pregnancy, the rate is per 10,000 live births and fetal deaths.

(c) Terminations of pregnancy <20 weeks gestation. TOP ≥ 20 weeks gestation or ≥ 400g birthweight are included as births.

Baby characteristics

Table 2.3.3: Births (gestation ≥20 weeks or birthweight ≥400g) with encephalocele by baby characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 births
Sex			
Male	27	40.9	0.5
Female	39	59.1	0.8
Gestational age (weeks)			
20–36	36	54.5	4.6
37–41	29	43.9	0.3
42 and over	1	1.5	0.5
Birthweight (grams)			
Less than 2,500	39	59.1	5.8
2,500–4,499	26	39.4	0.3
Not stated	1	1.5	. .

Maternal characteristics

Table 2.3.4: Number of women who gave birth to babies with encephalocele, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Women who gave birth	17	0.7	17	0.7	15	0.6	17	0.7	66	0.7

(a) The rate is per 10,000 women who gave birth.

Table 2.3.5: Number of women who gave birth to babies with encephalocele or who had a TOP where a fetus had encephalocele and the estimated rate, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Women who gave birth[(c)]	11	0.6	14	0.7	10	0.5	13	0.7	48	0.6
Women who gave birth or had a TOP[(d)]	19	1.0	24	1.3	29	1.5	21	1.1	93	1.2

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) The rate is per 10,000 women who gave birth.

(c) Includes women who had a termination of pregnancy at ≥ 20 weeks gestation or ≥ 400g birthweight.

(d) Women who had a termination of pregnancy at <20 weeks gestation.

Table 2.3.6: Births (gestation ≥ 20 weeks or birthweight ≥ 400g) with encephalocele by maternal characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 women
Maternal age group			
Less than 20	3	4.5	0.6
20–24	19	28.8	1.2
25–29	17	25.8	0.5
30–34	19	28.8	0.6
35–39	7	10.6	0.5
40 and over	1	1.5	0.4
Indigenous status[1]			
Indigenous	2	3.2	0.7
Non-Indigenous	61	96.8	0.6
Remoteness Area			
Major cities	47	71.2	0.8
Regional	16	24.2	0.6
Remote	2	3.0	0.9
Not stated	1	1.5	. .
Country of birth			
Australia	47	71.2	0.8
New Zealand	2	3.0	1.4
United Kingdom	2	3.0	0.6
Middle East and North Africa	3	4.5	1.6
Asia	7	10.6	1.0
Africa (excluding North Africa)	2	3.0	2.7
Not stated	3	4.5	0.1
Parity[2]			
Primiparous	16	29.1	0.6
Multiparous	38	69.1	1.0
Not stated	1	1.8	. .
Plurality			
Singleton	62	93.9	0.6
Multiple	4	6.1	2.5

(1) Excludes data from Tasmania

(2) Excludes data from New South Wales

Neural tube defects

This section includes all babies with anencephaly, spina bifida and encephalocele.

ICD-9-BPA codes: 740.00–742.09

For all births, the neural tube defect rate was 4.7 per 10,000 births (Table 2.4.1). The rate observed in the four states providing data on terminations of pregnancy <20 weeks was 2.5 times higher than the rate at birth (11.3 per 10,000 births) indicating a high rate of early terminations of pregnancies (Table 2.4.2).

The rate of neural tube defects was similar in males and females. The mean gestational age at birth was 30 completed weeks. The mean birthweight was 1,876g and 57.3% had birthweights below 2,500g (Table 2.4.3).

The mean maternal age for women who had a baby with a neural tube defect was 27.9 years. More younger women <25 years of age and older women ≥40years of age had given birth to a baby with a neural tube defect than women in the age group 25 to 39 years. Having an affected birth was three times higher in Indigenous women compared to non-Indigenous women. Women living in remote areas and multiparous women had higher rates of affected births. Women who had multiple births had a six times higher rate of an affected birth than the singletons (Table 2.4.6).

Reported rates of Neural tube defects

Table 2.4.1: Number and rate of neural tube defects by outcome, gestation ≥20 weeks or birthweight ≥400g, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Live births	73	2.9	72	2.9	74	2.9	58	2.3	277	2.8
Fetal deaths	54	31.3	46	26.4	45	25.4	53	30.7	198	28.4
All births	127	5.0	118	4.6	119	4.7	111	4.4	475	4.7

(a) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths.

Table 2.4.2: Number and the estimated rate of neural tube defects by outcome, gestation ≥20 weeks or birthweight ≥400g and TOP <20 weeks, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Live births	51	2.7	50	2.6	54	2.8	46	2.4	201	2.6
Fetal deaths	45	35.0	40	30.8	39	29.5	49	38.2	173	33.3
All births	96	5.0	90	4.6	93	4.8	95	5.0	374	4.8
Births and TOP[(c)]	219	11.4	230	11.8	220	11.4	203	10.6	872	11.3

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths. For births and terminations of pregnancy, the rate is per 10,000 live births and fetal deaths.

(c) Terminations of pregnancy <20 weeks gestation. TOP ≥ 20 weeks gestation or ≥ 400g birthweight are included as births.

Baby characteristics

Table 2.4.3: Births (gestation ≥ 20 weeks or birthweight ≥ 400g) with neural tube defects by baby characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 births
Sex			
Male	243	51.2	4.7
Female	226	47.6	4.6
Indeterminate	3	0.6	. .
Not stated	3	0.6	. .
Gestational age (weeks)			
20–36	273	57.5	35.3
37–41	193	40.6	2.1
42 and over	8	1.7	4.4
Not stated	1	0.2	. .
Birthweight (grams)			
Less than 2,500	272	57.3	40.5
2,500–4,499	183	38.5	2.0
4,500 and over	5	1.1	2.7
Not stated	15	3.2	. .

Maternal characteristics

Table 2.4.4: Number of women who gave birth to babies with neural tube defects, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Women who gave birth	127	5.1	118	4.7	119	4.8	111	4.5	475	4.8

(a) The rate is per 10,000 women who gave birth.

Table 2.4.5: Number of women who gave birth to babies with neural tube defects or who had a TOP where a fetus had neural tube defects and the estimated rate, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Women who gave birth[(c)]	96	5.1	90	4.7	93	4.9	95	5.1	374	4.9
Women who gave birth or had a TOP[(d)]	218	11.5	228	11.9	220	11.6	203	10.8	869	11.5

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) The rate is per 10,000 women who gave birth.

(c) Includes women who had a termination of pregnancy at ≥ 20 weeks gestation or ≥ 400g birthweight.

(d) Women who had a termination of pregnancy at <20 weeks gestation.

Table 2.4.6: Births (gestation ≥ 20 weeks or birthweight ≥ 400g) with neural tube defects by maternal characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 women
Maternal age group			
Less than 20	35	7.4	7.2
20–24	107	22.5	6.8
25–29	146	30.7	4.7
30–34	110	23.2	3.6
35–39	58	12.2	4.1
40 and over	14	2.9	5.4
Not stated	5	1.1	. .
Indigenous status[1]			
Indigenous	36	7.8	12.4
Non-Indigenous	414	89.4	4.4
Not stated	13	2.8	. .
Remoteness Area			
Major cities	297	62.5	4.8
Regional	136	28.6	5.3
Remote	25	5.3	11.1
Not stated	17	3.6	. .
Country of birth			
Australia	371	78.1	6.1
New Zealand	12	2.5	8.5
United Kingdom	15	3.2	4.6
Europe	7	1.5	3.1
Middle East and North Africa	14	2.9	7.5
Asia	26	5.5	3.7
North America	1	0.2	2.0
Africa (excluding North Africa)	3	0.6	4.1
Other countries	2	0.4	2.2
Not stated	24	5.1	. .
Parity[2]			
Primiparous	112	33.5	4.2
Multiparous	212	63.5	5.5
Not stated	10	3.0	. .
Plurality			
Singleton	429	90.3	4.4
Multiple	46	9.7	29.2

(1) Excludes data from Tasmania

(2) Excludes data from New South Wales

Microcephaly

Definition: A congenitally small cranium, defined by an occipito-frontal circumference (OFC) three standard deviations below the age and sex-appropriate distribution curves. Exclude microcephaly associated with anencephaly or encephalocele.

ICD-9-BPA codes: 742.1

Microcephaly can be present at birth or it may develop in the first few years of life. Babies born with microcephaly may have a smaller than normal head that will fail to grow as they progress through infancy. The lower rates seen in later years of this report could be due to delayed identification. About 2.2 per 10,000 births had microcephaly diagnosed over the four years (Table 2.5.1). The rate of babies born with microcephaly was almost equal among both sexes (49.3 male). Two thirds of the babies were born at term (mean gestational age was 35 completed weeks). The mean birthweight for those babies was 2,388g and a half had birthweight less than 2,500g (Table 2.5.3).

The average age of the women who had a pregnancy affected with microcephaly was 29.2 years. The rate of microcephaly births for Indigenous women was more than three times that observed for non-Indigenous women. Women living in remote areas had a three times greater rate than for women living in cities. Multiple births had a higher rate of babies with this anomaly compared to singleton births (Table 2.5.6).

Reported rates of Microcephaly

Table 2.5.1: Number and rate of microcephaly by outcome, gestation ≥20 weeks or birthweight ≥400g, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Live births	60	2.4	50	2.0	42	1.7	41	1.6	193	1.9
Fetal deaths	6	3.5	5	2.9	7	3.9	8	4.6	26	3.7
All births	66	2.6	55	2.2	49	1.9	49	2.0	219	2.2

(a) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths.

Table 2.5.2: Number and the estimated rate of microcephaly by outcome, gestation ≥20 weeks or birthweight ≥400g and TOP <20 weeks, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Live births	55	2.9	44	2.3	36	1.9	37	2.0	172	2.2
Fetal deaths	5	3.9	4	3.1	6	4.5	6	4.7	21	4.0
All births	60	3.1	48	2.5	42	2.2	43	2.3	193	2.5
Births and TOP[(c)]	63	3.3	50	2.6	46	2.4	49	2.6	208	2.7

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths. For births and terminations of pregnancy, the rate is per 10,000 live births and fetal deaths.

(c) Terminations of pregnancy <20 weeks gestation. TOP ≥ 20 weeks gestation or ≥ 400g birthweight are included as births.

Baby characteristics

Table 2.5.3: Births (gestation ≥20 weeks or birthweight ≥400g) with microcephaly by baby characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 births
Sex			
Male	108	49.3	2.1
Female	111	50.7	2.3
Gestational age (weeks)			
20–36	75	34.2	9.7
37–41	142	64.8	1.6
42 and over	1	0.5	0.5
Not stated	1	0.5	. .
Birthweight (grams)			
Less than 2,500	105	47.9	15.6
2,500–4,499	112	51.1	1.2
Not stated	2	0.9	. .

Maternal characteristics

Table 2.5.4: Number of women who gave birth to babies with microcephaly, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Women who gave birth	66	2.7	55	2.2	49	2.0	49	2.0	219	2.2

(a) The rate is per 10,000 women who gave birth.

Table 2.5.5: Number of women who gave birth to babies with microcephaly or who had a TOP where a fetus had microcephaly and the estimated rate, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Women who gave birth[(c)]	60	3.2	48	2.5	42	2.2	43	2.3	193	2.5
Women who gave birth or had a TOP[(d)]	63	3.3	50	2.6	46	2.4	49	2.6	208	2.7

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) The rate is per 10,000 women who gave birth.

(c) Includes women who had a termination of pregnancy at ≥ 20 weeks gestation or ≥ 400g birthweight.

(d) Women who had a termination of pregnancy at <20 weeks gestation.

Table 2.5.6: Births (gestation ≥ 20 weeks or birthweight ≥ 400g) with microcephaly by maternal characteristics, Australia, 1998–2001

Characteristic	Number	Percent	Rate per 10,000 women
Maternal age group			
Less than 20	10	4.6	2.1
20–24	32	14.6	2.0
25–29	66	30.1	2.1
30–34	74	33.8	2.4
35–39	26	11.9	1.8
40 and over	7	3.2	2.7
Not stated	4	1.8	. .
Indigenous status[(1)]			
Indigenous	19	8.7	6.5
Non-Indigenous	198	90.8	2.1
Not stated	1	0.5	. .
Remoteness Area			
Major cities	145	66.2	2.4
Regional	54	24.7	2.1
Remote	18	8.2	8.0
Not stated	2	0.9	. .
Country of birth			
Australia	163	74.4	2.7
New Zealand	2	0.9	1.4
United Kingdom	3	1.4	0.9
Europe	3	1.4	1.3
Middle East and North Africa	12	5.5	6.5
Asia	15	6.8	2.1
South and Central America and the Caribbean	3	1.4	5.9
Africa (excluding North Africa)	5	2.3	6.9
Other countries	2	0.9	2.2
Not stated	11	5.0	. .
Parity[(2)]			
Primiparous	71	39.9	2.7
Multiparous	100	56.2	2.6
Not stated	7	3.9	. .
Plurality			
Singleton	212	96.8	2.2
Multiple	7	3.2	4.4

(1) Excludes data from Tasmania

(2) Excludes data from New South Wales

Arhinencephaly/ Holoprosencephaly

Definition: A congenital anomaly of the brain, characterised by various degrees of incomplete lobation of the brain hemispheres. Olfactory nerve tract may be absent. Holoprosencephaly includes cyclopia, ethmocephaly, cebocephaly and premaxillary agenesis.

ICD-9-BPA codes: 742.26

ICD-10-AM codes: Q04.1, Q04.2 (ACT only)

The overall rate of arhinencephaly/holoprosencephaly at birth was about 0.7 per 10,000 births (Table 2.6.1). The rate of births and induced abortions from the four selected states was 1.4 per 10,000 births (Table 2.6.2).

There were more females than males with this anomaly. The mean gestational age at birth was 29 completed weeks. The mean birthweight was 1,477g and more than three-quarters of the babies weighed less than 2,500g (Table 2.6.3).

The mean maternal age of women who had a birth with arhinencephaly/holoprosencephaly was 27.1 years and this anomaly was more common among younger women who gave birth. Indigenous women had a higher rate of births with this anomaly than non-Indigenous women. The rate of this condition was four times greater for women who lived in remote areas than for women living in cities (Table 2.6.6).

Reported rates of arhinencephaly/holoprosencephaly

Table 2.6.1: Number and rate of arhinencephaly/holoprosencephaly by outcome, gestation ≥20 weeks or birthweight ≥400g, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Live births	8	0.3	8	0.3	14	0.6	13	0.5	43	0.4
Fetal deaths	5	2.9	7	4.0	11	6.2	8	4.6	31	4.4
All births	13	0.5	15	0.6	25	1.0	21	0.8	74	0.7

(a) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths.

Table 2.6.2: Number and the estimated rate of arhinencephaly/holoprosencephaly by outcome, gestation ≥20 weeks or birthweight ≥400g and TOP <20 weeks, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Live births	8	0.4	7	0.4	11	0.6	11	0.6	37	0.5
Fetal deaths	5	3.9	6	4.6	10	7.6	6	4.7	27	5.2
All births	13	0.7	13	0.7	21	1.1	17	0.9	64	0.8
Births and TOP[(c)]	24	1.2	31	1.6	31	1.6	23	1.2	109	1.4

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths. For births and terminations of pregnancy, the rate is per 10,000 live births and fetal deaths.

(c) Terminations of pregnancy <20 weeks gestation. TOP ≥ 20 weeks gestation or ≥ 400g birthweight are included as births

Baby characteristics

Table 2.6.3: Births (gestation ≥20 weeks or birthweight ≥400g) with arhinencephaly / holoprosencephaly by baby characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 births
Sex			
Male	32	43.2	0.6
Female	42	56.8	0.9
Gestational age (weeks)			
20–36	55	74.3	7.1
37–41	19	25.7	0.2
Birthweight (grams)			
Less than 2,500	57	77.0	8.5
2,500–4,499	17	23.0	0.2

Maternal characteristics

Table 2.6.4: Number of women who gave birth to babies with arhinencephaly/holoprosencephaly, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Women who gave birth	13	0.5	15	0.6	25	1.0	21	0.9	74	0.7

(a) The rate is per 10,000 women who gave birth.

Table 2.6.5: Number of women who gave birth to babies with arhinencephaly/holoprosencephaly or who had a TOP where a fetus had this anomaly and the estimated rate, available for four states[(a)] only,1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Women who gave birth[(c)]	13	0.7	13	0.7	21	1.1	17	0.9	64	0.8
Women who gave birth or had a TOP[(d)]	24	1.3	30	1.6	31	1.6	23	1.2	108	1.4

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) The rate is per 10,000 women who gave birth.

(c) Includes women who had a termination of pregnancy at ≥ 20 weeks gestation or ≥ 400g birthweight.

(d) Women who had a termination of pregnancy at <20 weeks gestation.

Table 2.6.6: Births (gestation ≥ 20 weeks or birthweight ≥ 400g) with arhinencephaly/ holoprosencephaly by maternal characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 women
Maternal age group			
Less than 20	12	16.2	2.5
20–24	12	16.2	0.8
25–29	24	32.4	0.8
30–34	19	25.7	0.6
35–39	6	8.1	0.4
40 and over	1	1.4	0.4
Indigenous status[(1)]			
Indigenous	6	8.2	2.1
Non-Indigenous	66	90.4	0.7
Not stated	1	1.4	. .
Remoteness Area			
Major cities	45	61.6	0.7
Regional	21	28.8	0.8
Remote	7	9.6	3.1
Country of birth			
Australia	56	75.7	0.9
New Zealand	1	1.4	0.7
United Kingdom	4	5.4	1.2
Europe	1	1.4	0.4
Middle East and North Africa	2	2.7	1.1
Asia	3	4.1	0.4
Africa (excluding North Africa)	3	4.1	4.1
Other countries	2	2.7	2.2
Not stated	2	2.7	. .
Parity[(2)]			
Primiparous	27	49.1	1.0
Multiparous	28	50.9	0.7
Plurality			
Singleton	70	94.6	0.7
Multiple	4	5.4	2.5

(1) Excludes data from Tasmania

(2) Excludes data from New South Wales

Hydrocephaly

Definition: A congenital anomaly characterised by dilatation of the cerebral ventricles, not associated with primary brain atrophy, with or without enlargement of the head, and diagnosed at birth. Not counted when present with encephalocele or spina bifida. Exclude: macrocephaly without dilatation of ventricular system, skull of macerated fetus, hydranencephaly, holoprosencephaly and postnatally acquired hydrocephalus.

ICD-9-BPA codes: 742.30–742.39

There were five cases of hydrocephalus per 10,000 births during the period 1998–2001 (Table 2.7.1). The data from the States providing data on termination of pregnancy before 20 weeks show that 7.3 cases per 10,000 births were affected with this anomaly (Table 2.7.2).

There were more males than females with hydrocephalus (58.1%) and the mean gestational age at birth was 31 completed weeks. The mean birthweight was 2,025g and 56.0% weighed less than 2,500g (Table 2.7.3).

The average age of women who had a baby with hydrocephaly was 28.9 years. Rates were higher for non-Indigenous women and multiparous women. This anomaly was four times higher for women having multiple births compared to singleton births (Table 2.7.6). Women living in remote areas were the least affected by a pregnancy with hydrocephalus.

Reported rates of hydrocephaly

Table 2.7.1: Number and rate of hydrocephaly by outcome, gestation ≥20 weeks or birthweight ≥400g, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Live births	83	3.3	91	3.6	89	3.5	90	3.6	353	3.5
Fetal deaths	32	18.5	41	23.5	46	25.9	31	17.9	150	21.5
All births	115	4.6	132	5.2	135	5.3	121	4.8	503	5.0

(a) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths.

Table 2.7.2: Number and the estimated rate of hydrocephaly by outcome, gestation ≥20 weeks or birthweight ≥400g and TOP <20 weeks, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Live births	71	3.7	74	3.8	73	3.8	74	3.9	292	3.8
Fetal deaths	30	23.3	36	27.7	40	30.2	30	23.4	136	26.2
All births	101	5.2	110	5.7	113	5.8	104	5.5	428	5.6
Births and TOP[(c)]	141	7.3	148	7.6	138	7.1	139	7.3	566	7.3

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths. For births and terminations of pregnancy, the rate is per 10,000 live births and fetal deaths.

(c) Terminations of pregnancy <20 weeks gestation. TOP ≥ 20 weeks gestation or ≥ 400g birthweight are included as births.

Baby characteristics

Table 2.7.3: Births (gestation ≥ 20 weeks or birthweight ≥ 400g) with hydrocephaly by baby characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 births
Sex			
Male	293	58.1	5.7
Female	205	40.7	4.2
Indeterminate	2	0.4	. .
Not stated	4	0.8	. .
Gestational age (weeks)			
20–36	301	59.7	38.9
37–41	195	38.7	2.1
42 and over	3	0.6	1.6
Not stated	5	1.0	. .
Birthweight (grams)			
Less than 2,500	282	56.0	42.0
2,500–4,499	204	40.5	2.2
4,500 and over	9	1.8	4.8
Not stated	9	1.8	. .

Maternal characteristics

Table 2.7.4: Number of women who gave birth to babies with hydrocephaly, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Women who gave birth	114	4.6	132	5.3	135	5.4	121	4.9	502	5.1

(a) The rate is per 10,000 women who gave birth.

Table 2.7.5: Number of women who gave birth to babies with hydrocephaly or who had a TOP where a fetus had this anomaly and the estimated rate, available for four states [(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Women who gave birth[(c)]	100	5.3	110	5.8	113	5.9	104	5.5	427	5.6
Women who gave birth or had a TOP[(d)]	140	7.4	148	7.7	138	7.2	139	7.4	565	7.4

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) The rate is per 10,000 women who gave birth.

(c) Includes women who had a termination of pregnancy at ≥ 20 weeks gestation or ≥ 400g birthweight.

(d) Women who had a termination of pregnancy at <20 weeks gestation.

Table 2.7.6: Births (gestation ≥ 20 weeks or birthweight ≥ 400g) with hydrocephaly by maternal characteristics, Australia, 1998–2001

Characteristic	Number	Percent	Rate per 10,000 women
Maternal age group			
Less than 20	35	7.0	7.2
20–24	80	15.9	5.1
25–29	148	29.4	4.7
30–34	138	27.4	4.5
35–39	85	16.9	6.0
40 and over	13	2.6	5.0
Not stated	4	0.8	. .
Indigenous status[1]			
Indigenous	11	2.2	3.8
Non-Indigenous	473	95.2	5.0
Not stated	13	2.6	. .
Remoteness Area			
Major cities	325	64.7	5.3
Regional	160	31.9	6.2
Remote	7	1.4	3.1
Not stated	10	2.0	. .
Country of birth			
Australia	372	74.0	6.1
New Zealand	10	2.0	7.1
United Kingdom	11	2.2	3.4
Europe	19	3.8	8.5
Middle East and North Africa	20	4.0	10.8
Asia	30	6.0	4.2
South and Central America and the Caribbean	3	0.6	5.9
Africa (excluding North Africa)	9	1.8	12.4
Other countries	11	2.2	10.9
Not stated	18	3.6	. .
Parity[2]			
Primiparous	159	41.0	6.0
Multiparous	229	59.0	5.9
Plurality			
Singleton	470	93.4	4.8
Multiple	33	6.6	20.9

(1) Excludes data from Tasmania

(2) Excludes data from New South Wales

Anophthalmos/ Microphthalmos

Definition: Apparently absent or small eyes. Some normal adnexal elements and eyelids are usually present. In microphthalmos, the corneal diameter is usually less than 10 mm and the antero-posterior diameter of the globe is less than 20 mm.

ICD-9-BPA codes: 743.0–743.1

Anophthalmos/microphthalmos was reported in 1 per 10,000 births (Table 2.8.1). Fetal deaths or terminations of pregnancies are not common for this anomaly.

More babies with this anomaly were male (57.5%). Most were born at term (67.9%) with a mean gestational age of 36 completed weeks. The mean birthweight for these babies were 2,721g and 63.2% had a birthweight ≥2,500g (Table 2.8.3).

The mean maternal age was 28.4 years. Women less than 20 years of age had the highest rate of anophthalmos/microphthalmos births. More women living in remote areas had a birth affected with this anomaly compared to the women living in cities or regional areas (Table 2.8.6).

Reported rates of anophthalmos/ microphthalmos

Table 2.8.1: Number and rate of anophthalmos/microphthalmos by outcome, gestation ≥20 weeks or birthweight ≥400g, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Live births	23	0.9	32	1.3	24	1.0	20	0.8	99	1.0
Fetal deaths	2	1.2	1	0.6	1	0.6	3	1.7	7	1.0
All births	25	1.0	33	1.3	25	1.0	23	0.9	106	1.0

(a) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths; the rate is per 1,000 fetal deaths.

Table 2.8.2: Number and the estimated rate of anophthalmos/microphthalmos by outcome, gestation ≥20 weeks or birthweight ≥400g and TOP <20 weeks, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Live births	22	1.1	27	1.4	20	1.0	12	0.6	81	1.1
Fetal deaths	2	1.6	1	0.8	1	0.8	2	1.6	6	1.2
All births	24	1.2	28	1.4	21	1.1	14	0.7	87	1.1
Births and TOP[(c)]	27	1.4	30	1.5	24	1.2	15	0.8	96	1.2

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths. For births and terminations of pregnancy, the rate is per 10,000 live births and fetal deaths.

(c) Terminations of pregnancy <20 weeks gestation. TOP ≥ 20 weeks gestation or ≥ 400g birthweight are included as births.

Baby characteristics

Table 2.8.3: Births (gestation ≥20 weeks or birthweight ≥400g) with anophthalmos/microphthalmos by baby characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 births
Sex			
Male	61	57.5	1.2
Female	44	41.5	0.9
Indeterminate	1	0.9	. .
Gestational age (weeks)			
20–36	32	30.2	4.1
37–41	72	67.9	0.8
42 and over	2	1.9	1.1
Birthweight (grams)			
Less than 2,500	39	36.8	5.8
2,500–4,499	65	61.3	0.7
4,500 and over	2	1.9	1.1

Maternal characteristics

Table 2.8.4: Number of women who gave birth to babies with anophthalmos/microphthalmos, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Women who gave birth	25	1.0	33	1.3	25	1.0	22	0.9	105	1.1

(a) The rate is per 10,000 women who gave birth.

Table 2.8.5: Number of women who gave birth to babies with anophthalmos/microphthalmos or who had a TOP where a fetus had this anomaly and the estimated rate, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Women who gave birth[(c)]	24	1.3	28	1.5	21	1.1	14	0.7	87	1.1
Women who gave birth or had a TOP[(d)]	27	1.4	30	1.6	24	1.3	15	0.8	96	1.3

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) The rate is per 10,000 women who gave birth.

(c) Includes women who had a termination of pregnancy at ≥ 20 weeks gestation or ≥ 400g birthweight.

(d) Women who had a termination of pregnancy at <20 weeks gestation.

Table 2.8.6: Births (gestation ≥ 20 weeks or birthweight ≥ 400g) with anophthalmos/microphthalmos by maternal characteristics, Australia, 1998–2001

Characteristic	Number	Percent	Rate per 10,000 women
Maternal age group			
Less than 20	11	10.5	2.3
20–24	13	12.4	0.8
25–29	34	32.4	1.1
30–34	31	29.5	1.0
35–39	12	11.4	0.8
40 and over	3	2.9	1.2
Not stated	1	1.0	. .
Indigenous status[1]			
Indigenous	5	4.8	1.7
Non-Indigenous	98	94.2	1.0
Not stated	1	1.0	. .
Remoteness Area			
Major cities	74	70.5	1.8
Regional	22	21.0	0.8
Remote	8	7.6	3.5
Not stated	1	1.0	. .
Country of birth			
Australia	74	70.5	1.2
New Zealand	2	1.9	1.4
United Kingdom	2	1.9	0.6
Europe	3	2.9	1.3
Middle East and North Africa	4	3.8	2.2
Asia	3	2.9	0.4
North America	0	0.0	0.0
South and Central America and the Caribbean	0	0.0	0.0
Africa (excluding North Africa)	5	4.8	6.9
Other countries	7	6.7	7.6
Not stated	5	4.8	. .
Parity[2]			
Primiparous	26	35.1	1.0
Multiparous	47	63.5	1.2
Not stated	1	1.4	. .
Plurality			
Singleton	99	94.3	1.0
Multiple	6	5.7	3.8

(1) Excludes data from Tasmania

(2) Excludes data from New South Wales

Microtia

Definition: A congenital anomaly characterised by absent parts of the pinna (with or without atresia of the ear canal) commonly expressed in grades (I–IV) of which the extreme form (grade IV) is anotia, absence of pinna. Exclude small, normally shaped ears, imperforate auditory meatus with a normal pinna, dysplastic and low set ears.

ICD-9-BPA codes: 744.21

ICD-10-AM codes: Q17.2 (ACT only)

There were 0.9 per 10,000 births affected with microtia each year (Table 2.9.1). Babies born with microtia were 50% male and female. The mean gestational age at birth was 38 weeks. The mean birthweight for these babies was 2,921g and 77.9% had birthweight $\geq$ 2,500g (Table 2.9.3).

The mean maternal age of women was 28.4 years. Indigenous women had a higher rate of babies with microtia than the non-Indigenous women (Table 2.9.6).

Reported rates of Microtia

Table 2.9.1: Number and rate of microtia by outcome, gestation ≥20 weeks or birthweight ≥400g, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Live births	21	0.8	17	0.7	18	0.7	25	1.0	81	0.8
Fetal deaths	2	1.2	0	0.0	3	1.7	0	0.0	5	0.7
All births	23	0.9	17	0.7	21	0.8	25	1.0	86	0.9

(a) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths.

Table 2.9.2: Number and the estimated rate of microtia by outcome, gestation ≥20 weeks or birthweight ≥400g and TOP <20 weeks, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Live births	19	1.0	16	0.8	14	0.7	21	1.1	70	0.9
Fetal deaths	2	1.6	0	0.0	3	2.3	0	0.0	5	1.0
All births	21	1.1	16	0.8	17	0.9	21	1.1	75	1.0
Births and TOP[(c)]	21	1.1	16	0.8	17	0.9	22	1.2	76	1.0

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths. For births and terminations of pregnancy, the rate is per 10,000 live births and fetal deaths.

(c) Terminations of pregnancy <20 weeks gestation. TOP ≥ 20 weeks gestation or ≥ 400g birthweight are included as births.

Baby characteristics

Table 2.9.3: Births (gestation ≥ 20 weeks or birthweight ≥ 400g) with microtia by baby characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 births
Sex			
Male	43	50.0	0.8
Female	43	50.0	0.9
Gestational age (weeks)			
20–36	15	17.4	1.9
37–41	67	77.9	0.7
42 and over	4	4.7	2.2
Birthweight (grams)			
Less than 2,500	19	22.1	2.8
2,500–4,499	67	77.9	0.7

Maternal characteristics

Table 2.9.4: Number of women who gave birth to babies with microtia, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Women who gave birth	23	0.9	17	0.7	21	0.8	25	1.0	86	0.9

(a) The rate is per 10,000 women who gave birth.

Table 2.9.5: Number of women who gave birth to babies with microtia or who had a TOP where a fetus had this anomaly and the estimated rate, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Women who gave birth[(c)]	21	1.1	16	0.8	17	0.9	21	1.1	75	1.0
Women who gave birth or had a TOP[(d)]	21	1.1	16	0.8	17	0.9	22	1.2	76	1.0

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) The rate is per 10,000 women who gave birth.

(c) Includes women who had a termination of pregnancy at ≥ 20 weeks gestation or ≥ 400g birthweight.

(d) Women who had a termination of pregnancy at <20 weeks gestation.

Table 2.9.6: Births (gestation ≥ 20 weeks or birthweight ≥ 400g) with microtia by maternal characteristics Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 women
Maternal age group			
Less than 20	6	7.0	1.2
20–24	18	20.9	1.2
25–29	26	30.2	0.8
30–34	20	23.3	0.6
35–39	13	15.1	0.9
40 and over	2	2.3	0.8
Not stated	1	1.2	. .
Indigenous status[1]			
Indigenous	4	4.7	1.4
Non-Indigenous	82	95.3	0.9
Remoteness Area			
Major cities	60	69.8	1.0
Regional	22	25.6	0.8
Remote	4	4.7	1.8
Country of birth			
Australia	62	72.1	1.0
New Zealand	2	2.3	1.4
United Kingdom	4	4.7	1.2
Middle East and North Africa	4	4.7	2.2
Asia	9	10.5	1.3
Africa (excluding North Africa)	1	1.2	1.4
Other countries	2	2.3	2.2
Not stated	2	2.3	. .
Parity[2]			
Primiparous	23	45.1	1.0
Multiparous	28	54.9	1.7
Plurality			
Singleton	84	97.7	0.9
Multiple	2	2.3	1.3

(1) Excludes data from Tasmania

(2) Excludes data from New South Wales

Transposition of great vessels

Definition: A cardiac defect where the aorta exits from the right ventricle and the pulmonary artery from the left ventricle, with or without other cardiac defects. Include double outlet ventricle so-called corrected transposition.

ICD-9-BPA codes: 745.10–745.19

About four babies per 10,000 births were diagnosed with transposition of great vessels during 1998 to 2001 (Table 2.10.1). The data from the four states that include TOP before 20 weeks show that there is a slight increase in the number of cases diagnosed from 1998 to 2001 (Table 2.10.2).

A higher proportion of babies with this anomaly were male (60.7%). The mean gestational age at birth was 37 completed weeks with most (82.5%) being born at term. The mean birthweight of babies born with transposition of great vessels was 3,033g and 81.3% had normal birthweight (Table 2.10.3).

The average maternal age of the women was 29.4 years and the highest rate was seen among women over 40 years of age. Women born in Africa had the highest rate (13.7 per 10,000 women who gave birth) of babies with this anomaly (Table 2.10.6). Women living in remote areas also had a higher rate of a birth with this anomaly.

Reported rates of transposition of great vessels

Table 2.10.1: Number and rate of transposition of great vessels by outcome, gestation ≥20 weeks or birthweight ≥400g, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Live births	95	3.8	94	3.7	82	3.3	99	4.0	370	3.7
Fetal deaths	11	6.4	9	5.2	10	5.6	2	1.2	32	4.6
All births	106	4.2	103	4.1	92	3.6	101	4.0	402	4.0

(a) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths.

Table 2.10.2: Number and the estimated rate of transposition of great vessels by outcome, gestation ≥20 weeks or birthweight ≥400g and TOP <20 weeks, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Live births	64	3.3	75	3.9	65	3.4	83	4.4	287	3.7
Fetal deaths	8	6.2	8	6.2	10	7.6	2	1.6	28	5.4
All births	72	3.7	83	4.3	75	3.9	85	4.5	315	4.1
Births and TOP[(c)]	76	3.9	88	4.5	80	4.1	92	4.8	336	4.4

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths. For births and terminations of pregnancy, the rate is per 10,000 live births and fetal deaths.

(c) Terminations of pregnancy <20 weeks gestation. TOP ≥ 20 weeks gestation or ≥ 400g birthweight are included as births.

Baby characteristics

Table 2.10.3: Births (gestation ≥20 weeks or birthweight ≥400g) with transposition of great vessels by baby characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 births
Sex			
Male	244	60.7	4.7
Female	157	39.1	3.2
Indeterminate	1	0.2	. .
Gestational age (weeks)			
20–36	70	17.4	9.0
37–41	327	81.3	3.6
42 and over	5	1.2	2.7
Birthweight (grams)			
Less than 2,500	74	18.4	11.0
2,500–4,499	316	78.6	3.4
4,500 and over	11	2.7	5.9
Not stated	1	0.2	. .

Maternal characteristics

Table 2.10.4: Number of women who gave birth to babies with transposition of great vessels, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Women who gave birth	106	4.3	103	4.1	92	3.7	101	4.1	402	4.0

(a) The rate is per 10,000 women who gave birth.

Table 2.10.5: Number of women who gave birth to babies with transposition of great vessels or who had a TOP where a fetus had this anomaly and the estimated rate, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Women who gave birth[(c)]	72	3.8	83	4.3	75	3.9	85	4.5	315	4.2
Women who gave birth or had a TOP[(d)]	76	4.0	88	4.6	80	4.2	92	4.9	336	4.4

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) The rate is per 10,000 women who gave birth.

(c) Includes women who had a termination of pregnancy at ≥ 20 weeks gestation or ≥ 400g birthweight.

(d) Women who had a termination of pregnancy at <20 weeks gestation.

Table 2.10.6: Births with transposition of great vessels (gestation ≥20 weeks or birthweight ≥400g) by maternal characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 women
Maternal age group			
Less than 20	16	4.0	3.3
20–24	64	15.9	4.1
25–29	121	30.1	3.9
30–34	124	30.8	4.0
35–39	58	14.4	4.1
40 and over	16	4.0	6.2
Not stated	3	0.7	. .
Indigenous status[1]			
Indigenous	15	3.7	5.2
Non-Indigenous	380	94.5	4.0
Not stated	7	1.7	. .
Remoteness Area			
Major cities	266	66.3	4.3
Regional	114	28.4	4.4
Remote	13	3.2	5.8
Not stated	8	2.0	. .
Country of birth			
Australia	302	75.1	5.0
New Zealand	10	2.5	7.1
United Kingdom	23	5.7	7.1
Europe	9	2.2	4.0
Middle East and North Africa	9	2.2	4.9
Asia	25	6.2	3.5
North America	2	0.5	4.1
South and Central America and the Caribbean	1	0.2	2.0
Africa (excluding North Africa)	10	2.5	13.7
Other countries	4	1.0	4.4
Not stated	7	1.7	. .
Parity[2]			
Primiparous	119	39.9	4.5
Multiparous	179	60.1	4.6
Plurality			
Singleton	391	97.3	4.0
Multiple	11	2.7	7.0

(1) Excludes data from Tasmania

(2) Excludes data from New South Wales

Tetralogy of Fallot

Definition: A condition characterised by ventricular septal defect, overriding aorta, infundibular pulmonary stenosis, and often right ventricular hypertrophy.

ICD-9-BPA codes: 745.20–745.21

There were 2.9 per 10,000 births affected with tetralogy of Fallot and this rate remained consistent across 1998 to 2001 (Table 2.11.1). The data provided by the states on termination of pregnancy before 20 weeks did not show a significant difference in the rate. (Table 2.11.2).

Nearly two-thirds of the babies with tetralogy of Fallot were male. 70.9% were born at term and the mean gestational age at birth was 36 completed weeks. The mean birthweight of those babies was 2,775g and more than two-thirds of them had birthweights above 2,500g.

The average age of women who gave birth to a baby with this anomaly was 30 years, and the anomaly was more common when the mother's age was 35 years or more. The rate of having a baby with this anomaly was higher for non-Indigenous women than for Indigenous women. This condition was more common among multiple births than in singletons (Table 2.11.6).

Reported rates of tetralogy of Fallot

Table 2.11.1: Number and rate of tetralogy of Fallot by outcome, gestation ≥20 weeks or birthweight ≥400g, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Live births	68	2.7	74	2.9	68	2.7	62	2.5	272	2.7
Fetal deaths	4	2.3	4	2.3	2	1.1	7	4.1	17	2.4
All births	72	2.9	78	3.1	70	2.8	69	2.8	289	2.9

(a) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths.

Table 2.11.2: Number and the estimated rate of tetralogy of Fallot by outcome, gestation ≥20 weeks or birthweight ≥400g and TOP <20 weeks, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Live births	52	2.7	50	2.6	57	3.0	43	2.3	202	2.6
Fetal deaths	4	3.1	4	3.1	2	1.5	4	3.1	14	2.7
All births	56	2.9	54	2.8	59	3.0	47	2.5	216	2.8
Births and TOP[(c)]	61	3.2	60	3.1	62	3.2	49	2.6	232	3.0

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths. For births and terminations of pregnancy, the rate is per 10,000 live births and fetal deaths.

(c) Terminations of pregnancy <20 weeks gestation. TOP ≥ 20 weeks gestation or ≥ 400g birthweight are included as births.

Baby characteristics

Table 2.11.3: Births (gestation ≥20 weeks or birthweight ≥400g) with tetralogy of Fallot by baby characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 births
Sex			
Male	184	63.7	3.5
Female	105	36.3	2.1
Gestational age (weeks)			
20–36	80	27.7	10.3
37–41	205	70.9	2.2
42 and over	4	1.4	2.2
Birthweight (grams)			
Less than 2,500	81	28.0	12.1
2,500–4,499	202	69.9	2.2
4,500 and over	5	1.7	2.7
Not stated	1	0.3	. .

Maternal characteristics

Table 2.11.4: Number of women who gave birth to babies with tetralogy of Fallot, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Women who gave birth	72	2.9	76	3.0	70	2.8	69	2.8	287	2.9

(a) The rate is per 10,000 women who gave birth.

Table 2.11.5: Number of women who gave birth to babies with tetralogy of Fallot or who had a TOP where a fetus had this anomaly and the estimated rate, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Women who gave birth[(c)]	56	3.0	53	2.8	59	3.1	47	2.5	215	2.8
Women who gave birth or had a TOP[(d)]	61	3.2	59	3.1	62	3.3	49	2.6	231	3.0

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) The rate is per 10,000 women who gave birth.

(c) Includes women who had a termination of pregnancy at ≥ 20 weeks gestation or ≥ 400g birthweight.

(d) Women who had a termination of pregnancy at <20 weeks gestation.

Table 2.11.6 Births (gestation ≥20 weeks or birthweight ≥400g) with tetralogy of Fallot by maternal characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 women
Maternal age group			
Less than 20	14	4.9	2.9
20–24	40	13.9	2.6
25–29	75	26.1	2.4
30–34	83	28.9	2.7
35–39	65	22.6	4.6
40 and over	10	3.5	3.9
Indigenous status[(1)]			
Indigenous	3	1.1	1.0
Non-Indigenous	277	97.5	2.9
Not stated	4	1.4	. .
Remoteness Area			
Major cities	202	70.4	3.3
Regional	75	26.1	2.9
Remote	5	1.7	2.2
Not stated	5	1.7	. .
Country of birth			
Australia	215	74.9	3.6
New Zealand	4	1.4	2.8
United Kingdom	5	1.7	1.5
Europe	12	4.2	5.4
Middle East and North Africa	9	3.1	4.9
Asia	26	9.1	3.7
North America	2	0.7	4.1
South and Central America and the Caribbean	1	0.3	2.0
Africa (excluding North Africa)	3	1.0	4.1
Other countries	5	1.7	5.5
Not stated	5	1.7	. .
Parity[(2)]			
Primiparous	98	43.0	3.7
Multiparous	129	56.6	3.3
Not stated	1	0.4	. .
Plurality			
Singleton	269	93.7	2.8
Multiple	18	6.3	11.4

(1) Excludes data from Tasmania

(2) Excludes data from New South Wales

Hypoplastic left heart syndrome

Definition: A cardiac defect with a hypoplastic left ventricle, associated with aortic and/or mitral valve atresia, with or without other cardiac defect.

ICD-9-BPA codes: 746.7

The hypoplastic left heart syndrome was diagnosed in 1.7 per 10,000 births. The data provided by four states that include termination of pregnancy before 20 weeks show that nearly half of affected pregnancies were diagnosed and terminated early in pregnancy or were still born (Table 2.12.2).

The condition was more common in males than in females. The mean gestational age at birth was 33 completed weeks and more than half were born at term. The mean birthweight of those babies was 2,247g (Table 2.12.3).

The average maternal age of women who had a baby with hypoplastic left heart syndrome was 28 years. More than 80% of women who had an affected baby were less than 35 years of age. Indigenous women had a higher rate of hypoplastic left heart syndrome births. Women who had a multiple births had a higher rate of affected births than the singleton births (Table 2.12.6).

Reported rates of Hypoplastic left heart syndrome

Table 2.12.1: Number and rate of hypoplastic left heart syndrome by outcome, gestation ≥20 weeks or birthweight ≥400g, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Live births	23	0.9	34	1.3	29	1.2	37	1.5	123	1.2
Fetal deaths	7	4.1	11	6.3	16	9.0	11	6.4	45	6.5
All births	30	1.2	45	1.8	45	1.8	48	1.9	168	1.7

(a) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths.

Table 2.12.2: Number and the estimated rate of hypoplastic left heart syndrome by outcome, gestation ≥20 weeks or birthweight ≥400g and TOP <20 weeks, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Live births	11	0.6	25	1.3	19	1.0	30	1.6	85	1.1
Fetal deaths	5	3.9	10	7.7	15	11.3	10	7.8	40	7.7
All births	16	0.8	35	1.8	34	1.8	40	2.1	125	1.6
Births and TOP[(c)]	24	1.2	44	2.3	43	2.2	44	2.3	155	2.0

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths. For births and terminations of pregnancy, the rate is per 10,000 live births and fetal deaths.

(c) Terminations of pregnancy <20 weeks gestation. TOP ≥ 20 weeks gestation or ≥ 400g birthweight are included as births.

Baby characteristics

Table 2.12.3: Births (gestation ≥20 weeks or birthweight ≥400g) with hypoplastic left heart syndrome by baby characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 births
Sex			
Male	109	64.9	2.1
Female	59	35.1	1.2
Gestational age (weeks)			
20–36	65	38.7	8.4
37–41	97	57.7	1.1
42 and over	3	1.8	1.6
Not stated	3	1.8	. .
Birthweight (grams)			
Less than 2,500	70	41.7	10.4
2,500–4,499	95	56.5	1.0
Not stated	3	1.8	. .

Maternal characteristics

Table 2.12.4: Number of women who gave birth to babies with hypoplastic left heart syndrome, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Women who gave birth	30	1.2	45	1.8	45	1.8	48	1.9	168	1.7

(a) The rate is per 10,000 women who gave birth.

Table 2.12.5: Number of women who gave birth to babies with hypoplastic left heart syndrome or who had a TOP where a fetus had this anomaly and the estimated rate, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Women who gave birth[(c)]	16	0.8	35	1.8	34	1.8	40	2.1	125	1.6
Women who gave birth or had a TOP[(d)]	24	1.3	44	2.3	43	2.3	44	2.3	155	2.0

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) The rate is per 10,000 women who gave birth.

(c) Includes women who had a termination of pregnancy at ≥ 20 weeks gestation or ≥ 400g birthweight.

(d) Women who had a termination of pregnancy at <20 weeks gestation.

Table 2.12.6: Births (gestation ≥20 weeks or birthweight ≥400g) with hypoplastic left heart by maternal characteristics, Australia, 1998–2001

Characteristic	Number	Percent	Rate per 10,000 women
Maternal age group			
Less than 20	9	5.4	1.9
20–24	40	23.8	2.6
25–29	51	30.4	1.6
30–34	35	20.8	1.1
35–39	27	16.1	1.9
40 and over	3	1.8	1.2
Not stated	3	1.8	. .
Indigenous status[1]			
Indigenous	7	4.2	2.4
Non-Indigenous	158	94.0	1.7
Not stated	3	1.8	. .
Remoteness Area			
Major cities	118	70.7	1.9
Regional	41	24.6	1.6
Remote	5	3.0	2.2
Not stated	3	1.8	. .
Country of birth			
Australia	128	76.2	2.1
New Zealand	4	2.4	2.8
United Kingdom	5	3.0	1.5
Europe	7	4.2	3.1
Middle East and North Africa	9	5.4	4.9
Asia	4	2.4	0.6
North America	2	1.2	4.1
South and Central America and the Caribbean	1	0.6	2.0
Other countries	4	2.4	4.4
Not stated	4	2.4	. .
Parity[2]			
Primiparous	57	41.3	2.2
Multiparous	80	58.0	2.1
Not stated	1	0.7	. .
Plurality			
Singleton	161	95.8	1.6
Multiple	7	4.2	4.4

(1) Excludes data from Tasmania

(2) Excludes data from New South Wales

Coarctation of the aorta

Definition: An obstruction in the descending aorta, almost invariably at the insertion of the ductus arteriosus.

ICD-9-BPA codes: 747.10–747.19

Coarctation of the aorta was reported in 3.1 per 10,000 births (Table 2.13.1). There were very few stillbirths or terminations of pregnancy reported for this condition (Table 2.13.2).

There were more males (59.0%) than females with coarctation of the aorta. The mean gestational age at birth was 37 completed weeks and most were born at term (79.4%). The mean birthweight for those babies was 2,962g and most (79.0%) weighed ≥2,500g (Table 2.13.3).

The average age of women who gave birth to a baby with this anomaly was 29.9 years. The rate of having an affected birth with this anomaly increased with advancing maternal age. This condition was more common among births of multiparous women compared to primiparous women. The rate of this anomaly was greater for women who had multiple births than for singleton births (Table 2.13.6).

Reported rates of coarctation of the aorta

Table 2.13.1: Number and rate of coarctation of aorta by outcome, gestation ≥20 weeks or birthweight ≥400g, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Live births	77	3.1	74	2.9	72	2.9	80	3.2	303	3.0
Fetal deaths	1	0.6	4	2.3	4	2.3	3	1.7	12	1.7
All births	78	3.1	78	3.1	76	3.0	83	3.3	315	3.1

(a) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths.

Table 2.13.2: Number and the estimated rate of coarctation of aorta by outcome, gestation ≥20 weeks or birthweight ≥400g and TOP <20 weeks, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Live births	58	3.0	59	3.1	59	3.1	65	3.4	241	3.1
Fetal deaths	1	0.8	4	3.1	4	3.0	2	1.6	11	2.1
All births	59	3.1	63	3.2	63	3.3	67	3.5	252	3.3
Births and TOP[(c)]	61	3.2	67	3.4	66	3.4	67	3.5	261	3.4

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths. For births and terminations of pregnancy, the rate is per 10,000 live births and fetal deaths.

(c) Terminations of pregnancy <20 weeks gestation. TOP ≥ 20 weeks gestation or ≥ 400g birthweight are included as births.

Baby characteristics

Table 2.13.3: Births (gestation ≥20 weeks or birthweight ≥400g) with coarctation of aorta by baby characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 births
Sex			
Male	186	59.0	3.6
Female	129	41.0	2.6
Gestational age (weeks)			
20–36	62	19.7	8.0
37–41	250	79.4	2.7
42 and over	1	0.3	0.5
Not stated	2	0.6	. .
Birthweight (grams)			
Less than 2,500	64	20.3	9.5
2,500–4,499	243	77.1	2.6
4,500 and over	6	1.9	3.2
Not stated	2	0.6	. .

Maternal characteristics

Table 2.13.4: Number of women who gave birth to babies with coarctation of aorta, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Women who gave birth	78	3.1	78	3.1	76	3.0	83	3.4	315	3.2

(a) The rate is per 10,000 women who gave birth.

Table 2.13.5: Number of women who gave birth to babies with coarctation of aorta or who had a TOP where a fetus had this anomaly and the estimated rate, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Women who gave birth[(c)]	59	3.1	63	3.3	63	3.3	67	3.6	252	3.3
Women who gave birth or had a TOP[(d)]	61	3.2	67	3.5	66	3.5	67	3.6	261	3.4

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) The rate is per 10,000 women who gave birth.

(c) Includes women who had a termination of pregnancy at ≥ 20 weeks gestation or ≥ 400g birthweight.

(d) Women who had a termination of pregnancy at <20 weeks gestation.

Table 2.13.6: Births (gestation ≥20 weeks or birthweight ≥400g) with coarctation of aorta by maternal characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 women
Maternal age group			
Less than 20	8	2.5	1.7
20–24	52	16.5	3.3
25–29	81	25.7	2.6
30–34	106	33.7	3.4
35–39	51	16.2	3.6
40 and over	11	3.5	4.3
Not stated	6	1.9	. .
Indigenous status[1]			
Indigenous	9	2.9	3.1
Non-Indigenous	300	95.5	3.2
Not stated	5	1.6	. .
Remoteness Area			
Major cities	213	67.6	3.5
Regional	94	29.8	3.6
Remote	7	2.2	3.1
Not stated	1	0.3	. .
Country of birth			
Australia	241	76.5	4.0
New Zealand	8	2.5	5.6
United Kingdom	14	4.4	4.3
Europe	8	2.5	3.6
Middle East and North Africa	10	3.2	5.4
Asia	15	4.8	2.1
North America	3	1.0	6.1
South and Central America and the Caribbean	2	0.6	4.0
Africa (excluding North Africa)	2	0.6	2.7
Other countries	3	1.0	3.3
Not stated	9	2.9	. .
Parity[2]			
Primiparous	87	34.9	3.3
Multiparous	162	65.1	4.2
Plurality			
Singleton	301	95.6	3.1
Multiple	14	4.4	8.9

(1) Excludes data from Tasmania

(2) Excludes data from New South Wales

Choanal atresia

Definition: Congenital obstruction (membraneous or osseous) of the posterior choana or choanae. Exclude choanal stenosis and congestion of nasal mucosa.

ICD-9-BPA codes: 748.0

The rate of choanal atresia in births remained fairly consistent during 1998 to 2001, with an overall rate of 1.2 per 10,000 births. Stillbirths or terminations of pregnancy due to this condition were rare (Table 2.14.1).

More females than males were affected (54.4%) with this anomaly. The mean gestational age at birth was 37 completed weeks and three-quarters of them (77.6%) were born at term. The mean birthweight was 2,921g and 76% weighed ≥2,500g (Table 2.14.3).

The average maternal age was 29.6 years (Table 2.14.6).

Reported rates of Choanal atresia

Table 2.14.1: Number and rate of choanal atresia by outcome, gestation ≥20 weeks or birthweight ≥400g, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Live births	36	1.4	29	1.2	27	1.1	30	1.2	122	1.2
Fetal deaths	0	0.0	0	0.0	2	1.1	1	0.6	3	0.4
All births	36	1.4	29	1.1	29	1.1	31	1.2	125	1.2

(a) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths.

Table 2.14.2: Number and the estimated rate of choanal atresia by outcome, gestation ≥20 weeks or birthweight ≥400g and TOP <20 weeks, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Live births	33	1.7	25	1.3	24	1.2	27	1.4	109	1.4
Fetal deaths	0	0.0	0	0.0	1	0.8	1	0.8	2	0.4
All births	33	1.7	25	1.3	25	1.3	28	1.5	111	1.4
Births and TOP[(c)]	33	1.7	26	1.3	25	1.3	28	1.5	112	1.5

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths. For births and terminations of pregnancy, the rate is per 10,000 live births and fetal deaths.

(c) Terminations of pregnancy <20 weeks gestation. TOP ≥ 20 weeks gestation or ≥ 400g birthweight are included as births.

Baby characteristics

Table 2.14.3: Births (gestation ≥20 weeks or birthweight ≥400g) with choanal atresia by baby characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 births
Sex			
Male	57	45.6	1.1
Female	68	54.4	1.4
Gestational age (weeks)			
20–36	27	21.6	3.5
37–41	97	77.6	1.1
42 and over	1	0.8	0.5
Birthweight (grams)			
Less than 2,500	29	23.2	4.3
2,500–4,499	94	75.2	1.0
4,500 and over	1	0.8	0.5
Not stated	1	0.8	. .

Maternal characteristics

Table 2.14.4: Number of women who gave birth to babies with choanal atresia, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Women who gave birth	36	1.5	29	1.2	29	1.2	31	1.3	125	1.3

(a) The rate is per 10,000 women who gave birth.

Table 2.14.5: Number of women who gave birth to babies with choanal atresia or who had a TOP where a fetus had this anomaly and the estimated rate, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Women who gave birth[(c)]	33	1.7	25	1.3	25	1.3	28	1.5	111	1.5
Women who gave birth or had a TOP[(d)]	33	1.7	26	1.4	25	1.3	28	1.5	112	1.5

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) The rate is per 10,000 women who gave birth.

(c) Includes women who had a termination of pregnancy at ≥ 20 weeks gestation or ≥ 400g birthweight.

(d) Women who had a termination of pregnancy at <20 weeks gestation.

Table 2.14.6: Births (gestation ≥20 weeks or birthweight ≥400g) with choanal atresia by maternal characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 women
Maternal age group			
Less than 20	5	4.0	1.0
20–24	14	11.2	0.9
25–29	43	34.4	1.4
30–34	39	31.2	1.3
35–39	20	16.0	1.4
40 and over	4	3.2	1.5
Indigenous status[(1)]			
Indigenous	3	2.4	1.0
Non-Indigenous	118	95.2	1.3
Not stated	3	2.4	. .
Remoteness Area			
Major cities	84	67.7	1.4
Regional	38	30.6	1.5
Remote	2	1.6	0.9
Country of birth			
Australia	105	84.0	1.7
New Zealand	2	1.6	1.4
United Kingdom	1	0.8	0.3
Europe	7	5.6	3.1
Middle East and North Africa	2	1.6	1.1
Asia	4	3.2	0.6
Africa (excluding North Africa)	1	0.8	1.4
Other countries	1	0.8	1.1
Not stated	2	1.6	. .
Parity[(2)]			
Primiparous	31	32.0	1.2
Multiparous	65	67.0	1.7
Not stated	1	1.0	. .
Plurality			
Singleton	119	95.2	1.2
Multiple	6	4.8	3.8

(1) Excludes data from Tasmania

(2) Excludes data from New South Wales

Cleft palate without cleft lip

Definition: A congenital anomaly characterised by a closure defect of the hard and/or soft palate behind the foramen incisivum without cleft lip. Include sub mucous cleft palate. Exclude cleft palate with cleft lip, cleft uvula, functional short palate, and high narrow palate.

ICD-9-BPA codes: 749.00–749.09

There were 7.6per 10,000 births with cleft palate without cleft lip and this rate has not changed during the period 1998–2001.

The proportion of females with this anomaly was 53.0%. The mean gestational age at birth was 37 completed weeks and 80.3% were born at term. The mean birthweight was 3,038g and most of them had birthweights ≥2,500g (Table 2.15.3).

The average maternal age was 29.3 years. The women born in New Zealand had the highest rate of an affected birth. A higher rate was observed for births of multiparous women and multiple births (Table 2.15.6).

Reported rates of cleft palate without cleft lip

Table 2.15.1: Number and rate of cleft palate without cleft lip by outcome, gestation ≥20 weeks or birthweight ≥400g, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Live births	188	7.5	173	6.9	174	6.9	189	7.6	724	7.2
Fetal deaths	10	5.8	10	5.7	12	6.8	11	6.4	43	6.2
All births	198	7.9	183	7.2	186	7.3	200	8.0	767	7.6

(a) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths.

Table 2.15.2: Number and the estimated rate of cleft palate without cleft lip by outcome, gestation ≥20 weeks or birthweight ≥400g and TOP <20 weeks, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Live births	145	7.6	140	7.3	153	8.0	151	8.0	589	7.7
Fetal deaths	8	6.2	8	6.2	11	8.3	11	8.6	38	7.3
All births	153	7.9	148	7.6	164	8.5	162	8.5	627	8.1
Births and TOP[(c)]	157	8.1	155	8.0	174	9.0	167	8.8	653	8.5

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths. For births and terminations of pregnancy, the rate is per 10,000 live births and fetal deaths.

(c) Terminations of pregnancy <20 weeks gestation. TOP ≥ 20 weeks gestation or ≥ 400g birthweight are included as births.

Baby characteristics

Table 2.15.3: Births (gestation ≥20 weeks or birthweight ≥400g) with cleft palate without cleft lip by baby characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 births
Sex			
Male	358	46.6	6.9
Female	407	53.0	8.3
Indeterminate	2	0.3	. .
Not stated	1	0.1	. .
Gestational age (weeks)			
20–36	130	16.9	16.8
37–41	617	80.3	6.8
42 and over	16	2.1	8.8
Not stated	5	0.7	. .
Birthweight (grams)			
Less than 2,500	145	18.9	21.6
2,500–4,499	602	78.4	6.5
4,500 and over	15	2.0	8.0
Not stated	6	0.8	. .

Maternal characteristics

Table 2.15.4: Number of women who gave birth to babies with cleft palate without cleft lip, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Women who gave birth	198	8.0	183	7.3	186	7.5	199	8.1	766	7.7

(a) The rate is per 10,000 women who gave birth.

Table 2.15.5: Number of women who gave birth to babies with cleft palate without cleft lip or who had a TOP where a fetus had this anomaly and the estimated rate, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Women who gave birth[(c)]	153	8.1	148	7.7	164	8.6	161	8.6	626	8.3
Women who gave birth or had a TOP[(d)]	157	8.3	155	8.1	174	9.1	166	8.9	652	8.6

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) The rate is per 10,000 women who gave birth.

(c) Includes women who had a termination of pregnancy at ≥ 20 weeks gestation or ≥ 400g birthweight.

(d) Women who had a termination of pregnancy at <20 weeks gestation.

Table 2.15.6: Births (gestation ≥20 weeks or birthweight ≥400g) with cleft palate without cleft lip by maternal characteristics, Australia, 1998–2001

Characteristic	Number	Percent	Rate per 10,000 women
Maternal age group			
Less than 20	35	4.6	7.2
20–24	120	15.6	7.7
25–29	250	32.6	8.0
30–34	208	27.1	6.7
35–39	123	16.0	8.7
40 and over	25	3.3	9.7
Not stated	6	0.8	. .
Indigenous status[1]			
Indigenous	28	3.7	9.6
Non-Indigenous	718	95.2	7.6
Not stated	8	1.1	. .
Remoteness Area			
Major cities	504	65.7	8.2
Regional	232	30.2	9.0
Remote	16	2.1	7.1
Not stated	15	2.0	. .
Country of birth			
Australia	586	76.4	9.7
New Zealand	24	3.1	16.9
United Kingdom	31	4.0	9.6
Europe	25	3.3	11.2
Middle East and North Africa	7	0.9	3.8
Asia	59	7.7	8.3
South and Central America and the Caribbean	3	0.4	5.9
Africa (excluding North Africa)	2	0.3	2.7
Other countries	10	1.3	10.9
Not stated	20	2.6	. .
Parity[2]			
Primiparous	195	36.5	7.4
Multiparous	335	62.7	8.6
Not stated	4	0.7	. .
Plurality			
Singleton	740	96.5	7.6
Multiple	27	3.5	17.1

(1) Excludes data from Tasmania

(2) Excludes data from New South Wales

Cleft lip with or without cleft palate

Definition: A congenital anomaly characterised by partial or complete clefting of the upper lip, with or without clefting of the alveolar ridge or the hard palate. Exclude midline cleft of upper or lower lip and oblique facial fissure (going towards the eye).

ICD-9-BPA codes: 749.10–749.19, 749.20–749.29

There were 9.5 cases of cleft lip with or without cleft palate per 10,000 births reported during the period 1998 to 2001.

This anomaly was more commonly seen in males than in females. The mean gestational age at birth was 37 completed weeks and 79.4% were born at term. The mean birthweight was 3,043g and 79.4% weighed ≥2,500g (Table 2.16.3).

The mean maternal age was 28.6 years. This condition was more common among the babies of Indigenous women than of non Indigenous women. It was also more common among multiple than singleton births (Table 2.16.6) and the babies of women lived in remote areas.

Reported rates of cleft lip with or without cleft palate

Table 2.16.1: Number and rate of cleft lip with or without cleft palate by outcome, gestation ≥20 weeks or birthweight ≥400g, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Live births	231	9.2	221	8.8	237	9.4	217	8.7	906	9.0
Fetal deaths	13	7.5	13	7.5	14	7.9	13	7.5	53	7.6
All births	244	9.7	234	9.2	251	9.9	230	9.2	959	9.5

(a) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths.

Table 2.16.2: Number and the estimated rate of cleft lip with or without cleft palate by outcome, gestation ≥20 weeks or birthweight ≥400g and TOP <20 weeks, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Live births	185	9.7	171	8.9	166	8.6	159	8.4	681	8.9
Fetal deaths	13	10.1	13	10.0	13	9.8	12	9.4	51	9.8
All births	198	10.3	184	9.5	179	9.2	171	9.0	732	9.5
Births and TOP[(c)]	219	11.4	203	10.5	199	10.3	188	9.9	809	10.5

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths. For births and terminations of pregnancy, the rate is per 10,000 live births and fetal deaths.

(c) Terminations of pregnancy <20 weeks gestation. TOP ≥ 20 weeks gestation or ≥ 400g birthweight are included as births.

Baby characteristics

Table 2.16.3: Births (gestation ≥20 weeks or birthweight ≥400g) with cleft lip with or without cleft palate by baby characteristics, Australia, 1998–2001

Characteristic	Number	Percent	Rate per 10,000 births
Sex			
Male	603	62.9	11.6
Female	355	37.0	7.2
Indeterminate	1	0.1	. .
Gestational age (weeks)			
20–36	177	18.5	22.9
37–41	761	79.4	8.3
42 and over	21	2.2	11.5
Birthweight (grams)			
Less than 2,500	197	20.5	29.3
2,500–4,499	748	78.0	8.1
4,500 and over	13	1.4	7.0
Not stated	1	0.1	. .

Maternal characteristics

Table 2.16.4: Number of women who gave birth to babies with cleft lip with or without cleft palate, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Women who gave birth	244	9.8	234	9.4	251	10.1	229	9.3	958	9.6

(a) The rate is per 10,000 women who gave birth.

Table 2.16.5: Number of women who gave birth to babies with cleft lip with or without cleft palate or who had a TOP where a fetus had this anomaly and the estimated rate, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Women who gave birth[(c)]	198	10.4	184	9.6	179	9.4	171	9.1	732	9.6
Women who gave birth or had a TOP[(d)]	219	11.5	203	10.6	199	10.5	188	10.0	809	10.7

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) The rate is per 10,000 women who gave birth.

(c) Includes women who had a termination of pregnancy at ≥ 20 weeks gestation or ≥ 400g birthweight.

(d) Women who had a termination of pregnancy at <20 weeks gestation.

Table 2.16.6: Births (gestation ≥20 weeks or birthweight ≥400g) with cleft lip with or without cleft palate by maternal characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 women
Maternal age group			
Less than 20	73	7.6	15.1
20–24	168	17.5	10.7
25–29	284	29.6	9.1
30–34	271	28.3	8.8
35–39	132	13.8	9.3
40 and over	29	3.0	11.2
Not stated	1	0.1	. .
Indigenous status[(1)]			
Indigenous	46	4.9	15.8
Non-Indigenous	886	94.4	9.4
Not stated	7	0.7	. .
Remoteness Area			
Major cities	587	61.4	9.5
Regional	311	32.5	12.0
Remote	33	3.5	14.6
Not stated	25	2.6	. .
Country of birth			
Australia	752	78.5	12.4
New Zealand	20	2.1	14.1
United Kingdom	46	4.8	14.2
Europe	19	2.0	8.5
Middle East and North Africa	6	0.6	3.2
Asia	65	6.8	9.1
North America	3	0.3	6.1
South and Central America and the Caribbean	8	0.8	15.9
Africa (excluding North Africa)	9	0.9	12.4
Other countries	16	1.7	17.4
Not stated	14	1.5	. .
Parity[(2)]			
Primiparous	244	36.5	9.2
Multiparous	424	63.4	10.9
Not stated	1	0.1	. .
Plurality			
Singleton	929	97.0	9.5
Multiple	29	3.0	18.4

(1) Excludes data from Tasmania

(2) Excludes data from New South Wales

Oesophageal atresia / stenosis

Definition: A congenital anomaly characterised by the absence of continuity or narrowing of the oesophagus, with or without tracheal fistula, including tracheoesophageal fistula with or without mention of atresia or stenosis of oesophagus.

ICD-9-BPA codes: 750.30– 750.38

There were 2.6 reported cases of oesophageal atresia per 10,000 births during 1998 to 2001. There were very few stillbirths or terminations of pregnancy reported.

More males than females were affected with this anomaly. The mean gestational age at birth was 35 completed weeks and more than half (57.1%) were born at term. The mean birthweight was 2,322g and half of those babies (52.1%) weighed less than 2,500g (Table 2.17.3).

The average age of women who gave birth to a baby with oesophageal atresia and stenosis was 29.4 years. There was no difference in rates between Indigenous and non-Indigenous women. This anomaly was more common in multiple births than in singletons (Table 2.17.6).

Reported rates of Oesophageal atresia/stenosis

Table 2.17.1: Number and rate of oesophageal atresia/stenosis by outcome, gestation ≥20 weeks or birthweight ≥400g, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Live births	67	2.7	59	2.3	59	2.3	53	2.1	238	2.4
Fetal deaths	2	1.2	6	3.4	6	3.4	7	4.1	21	3.0
All births	69	2.7	65	2.6	65	2.6	60	2.4	259	2.6

(a) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths.

Table 2.17.2: Number and the estimated rate of oesophageal atresia/stenosis by outcome, gestation ≥20 weeks or birthweight ≥400g and TOP <20 weeks, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Live births	51	2.7	51	2.6	46	2.4	40	2.1	188	2.5
Fetal deaths	2	1.6	5	3.8	4	3.0	6	4.7	17	3.3
All births	53	2.8	56	2.9	50	2.6	46	2.4	205	2.7
Births and TOP[(c)]	55	2.9	62	3.2	54	2.8	48	2.5	219	2.8

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths. For births and terminations of pregnancy, the rate is per 10,000 live births and fetal deaths.

(c) Terminations of pregnancy <20 weeks gestation. TOP ≥ 20 weeks gestation or ≥ 400g birthweight are included as births.

Baby characteristics

Table 2.17.3: Births (gestation ≥20 weeks or birthweight ≥400g) with oesophageal atresia/stenosis by baby characteristics, Australia, 1998–2001

Characteristic	Number	Perent	Rate per 10,000 births
Sex			
Male	147	56.8	2.8
Female	112	43.2	2.3
Gestational age (weeks)			
20–36	109	42.1	14.1
37–41	148	57.1	1.6
42 and over	2	0.8	1.1
Birthweight (grams)			
Less than 2,500	135	52.1	20.1
2,500–4,499	123	47.5	1.3
Not stated	1	0.4	. .

Maternal characteristics

Table 2.17.4: Number of women who gave birth to babies with oesophageal atresia / stenosis, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Women who gave birth	69	2.8	64	2.6	65	2.6	60	2.4	258	2.6

(a) The rate is per 10,000 women who gave birth.

Table 2.17.5: Number of women who gave birth to babies with oesophageal atresia/ stenosis or who had a TOP where a fetus had this anomaly and the estimated rate, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Women who gave birth[(c)]	53	2.8	55	2.9	50	2.6	46	2.5	204	2.7
Women who gave birth or had a TOP[(d)]	55	2.9	61	3.2	54	2.8	48	2.6	218	2.9

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) The rate is per 10,000 women who gave birth.

(c) Includes women who had a termination of pregnancy at ≥ 20 weeks gestation or ≥ 400g birthweight.

(d) Women who had a termination of pregnancy at <20 weeks gestation.

Table 2.17.6: Births (gestation ≥20 weeks or birthweight ≥400g) with oesophageal atresia/stenosis by maternal characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 women
Maternal age group			
Less than 20	15	5.8	3.1
20–24	38	14.7	2.4
25–29	74	28.7	2.4
30–34	80	31.0	2.6
35–39	45	17.4	3.2
40 and over	6	2.3	2.3
Indigenous status[1]			
Aboriginal or Torres Strait Islander	8	3.1	2.8
Non-Indigenous	244	96.1	2.6
Not stated	2	0.8	. .
Remoteness Area			
Major cities	182	70.5	3.0
Regional	62	24.0	2.4
Remote	8	3.1	3.5
Not stated	6	2.3	. .
Country of birth			
Australia	205	79.5	3.4
New Zealand	4	1.6	2.8
United Kingdom	9	3.5	2.8
Europe	12	4.7	5.4
Middle East and North Africa	8	3.1	4.3
Asia	11	4.3	1.5
North America	1	0.4	2.0
South and Central America and the Caribbean	4	1.6	7.9
Africa (excluding North Africa)	1	0.4	1.4
Other countries	1	0.4	1.1
Not stated	2	0.8	. .
Parity[2]			
Primiparous	86	48.3	3.3
Multiparous	92	51.7	2.4
Plurality			
Singleton	238	92.2	2.4
Multiple	20	7.8	12.7

(1) Excludes data from Tasmania

(2) Excludes data from New South Wales

Small intestine atresia/stenosis

Definition: Complete or partial occlusion of the lumen of a segment of the small intestine. It can involve a single area or multiples areas of the jejunum or ileum. Exclude duodenal atresia.

ICD-9-BPA codes: 751.10–751.19

There were 2.4 reported cases of small intestinal atresia/stenosis per 10,000 births during the period 1998 to 2001. Stillbirths or terminations of pregnancy were not common for small intestinal atresia/stenosis (Table 2.18.1).

The mean gestational age at birth was 35 completed weeks and half of those babies (52.2%) were born preterm. This condition was more common among female births. The mean birthweight was 2,567g (Table 2.18.3).

The mean maternal age of women who gave birth to a baby with intestinal atresia/stenosis was 29 years. Women aged less than 20 years and the women aged more than 40 years had the highest rate of babies affected with this anomaly. More non-Indigenous women had babies with this anomaly than Indigenous women. This condition was less common among babies born to women lived in remote areas. The rate of having a baby with this anomaly was three times higher for multiple births than for singleton births (Table 2.18.6).

Reported rates Small intestine atresia/stenosis

Table 2.18.1: Number and rate of small intestine atresia/stenosis by outcome, gestation ≥20 weeks or birthweight ≥400g, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Live births	51	2.0	66	2.6	58	2.3	58	2.3	233	2.3
Fetal deaths	1	0.6	2	1.1	5	2.8	4	2.3	12	1.7
All births	52	2.1	68	2.7	63	2.5	62	2.5	245	2.4

(a) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths.

Table 2.18.2: Number and the estimated rate of small intestine atresia/stenosis by outcome, gestation ≥20 weeks or birthweight ≥400g and TOP <20 weeks, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Live births	40	2.1	53	2.7	50	2.6	47	2.5	190	2.5
Fetal deaths	1	0.8	1	0.8	4	3.0	4	3.1	10	1.9
All births	41	2.1	54	2.8	54	2.8	51	2.7	200	2.6
Births and TOP[(c)]	41	2.1	56	2.9	56	2.9	52	2.7	205	2.7

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths. For births and terminations of pregnancy, the rate is per 10,000 live births and fetal deaths.

(c) Terminations of pregnancy <20 weeks gestation. TOP ≥ 20 weeks gestation or ≥ 400g birthweight are included as births.

Baby characteristics

Table 2.18.3: Births (gestation ≥20 weeks or birthweight ≥400g) with small intestine atresia/stenosis by baby characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 births
Sex			
Male	113	46.1	2.2
Female	132	53.9	2.7
Gestational age (weeks)			
20–36	128	52.2	16.5
37–41	117	47.8	1.3
Birthweight (grams)			
Less than 2,500	109	44.5	16.2
2,500–4,499	133	54.3	1.4
4,500 and over	2	0.8	1.1
Not stated	1	0.4	. .

Maternal characteristics

Table 2.18.4: Number of women who gave birth to babies with small intestine atresia/stenosis, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Women who gave birth	52	2.1	68	2.7	63	2.5	62	2.5	245	2.5

(a) The rate is per 10,000 women who gave birth.

Table 2.18.5: Number of women who gave birth to babies with small intestine atresia/stenosis or who had a TOP where a fetus had this anomaly and the estimated rate, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Women who gave birth[(c)]	41	2.2	54	2.8	54	2.8	51	2.7	200	2.6
Women who gave birth or had a TOP[(d)]	41	2.2	56	2.9	56	2.9	52	2.8	205	2.7

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) The rate is per 10,000 women who gave birth.

(c) Includes women who had a termination of pregnancy at ≥ 20 weeks gestation or ≥ 400g birthweight

(d) Women who had a termination of pregnancy at <20 weeks gestation.

Table 2.18.6: Births (gestation ≥20 weeks or birthweight ≥400g) with small intestine atresia/stenosis by maternal characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 women
Maternal age group			
Less than 20	20	8.2	4.1
20–24	44	18.0	2.8
25–29	66	26.9	2.1
30–34	66	26.9	2.1
35–39	33	13.5	2.3
40 and over	14	5.7	5.4
Not stated	2	0.8	. .
Indigenous status[(1)]			
Indigenous	4	1.7	1.4
Non-Indigenous	231	95.9	2.5
Not stated	6	2.5	. .
Remoteness Area			
Major cities	158	64.8	2.6
Regional	78	32.0	3.0
Remote	3	1.2	1.3
Not stated	5	2.0	. .
Country of birth			
Australia	181	73.9	3.0
New Zealand	4	1.6	2.8
United Kingdom	16	6.5	4.9
Europe	10	4.1	4.5
Middle East and North Africa	8	3.3	4.3
Asia	17	6.9	2.4
North America	2	0.8	4.1
South and Central America and the Caribbean	0	0.0	0.0
Africa (excluding North Africa)	1	0.4	1.4
Other countries	2	0.8	2.2
Not stated	4	1.6	. .
Parity[(2)]			
Primiparous	73	42.4	2.8
Multiparous	98	57.0	2.5
Not stated	1	0.6	. .
Plurality			
Singleton	231	94.3	2.4
Multiple	14	5.7	8.9

(1) Excludes data from Tasmania

(2) Excludes data from New South Wales

Anorectal atresia/stenosis

Definition: A congenital anomaly characterised by absence of continuity of the anorectal canal or of communication between rectum and anus, or narrowing of anal canal, with or without fistula to neighbouring organs. Exclude mild stenosis which does not need correction, and ectopic anus.

ICD-9-BPA codes: 751.20–751.24

Anorectal atresia/stenosis was diagnosed in 3.3 per 10,000 births. The data provided by four states that include termination of pregnancy before 20 weeks show a higher rate for this condition. However, this rate slightly decreased from 1998 to 2001 (Table 2.19.2).

There were more males than females born with this anomaly. The mean gestational age at birth was 35 completed weeks and 64.4% of the babies were born at term. The mean birthweight was 2,589g (Table 2.19.3).

The mean age of women who had a baby with anorectal atresia/stenosis was 28.5years. Indigenous women had a higher rate of births with this anomaly compared to non-Indigenous women. Women living in remote areas and multiple births also had a higher rate of births with this anomaly (Table 2.19.6).

Reported rates of anorectal atresia/stenosis

Table 2.19.1: Number and rate of anorectal atresia/stenosis by outcome, gestation ≥20 weeks or birthweight ≥400g, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Live births	78	3.1	68	2.7	77	3.1	67	2.7	290	2.9
Fetal deaths	14	8.1	9	5.2	5	2.8	13	7.5	41	5.9
All births	92	3.7	77	3.0	82	3.2	80	3.2	331	3.3

(a) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths.

Table 2.19.2: Number and the estimated rate of anorectal atresia/stenosis by outcome, gestation ≥20 weeks or birthweight ≥400g and TOP <20 weeks, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Live births	63	3.3	60	3.1	58	3.0	50	2.6	231	3.0
Fetal deaths	13	10.1	7	5.4	4	3.0	12	9.4	36	6.9
All births	76	3.9	67	3.4	62	3.2	62	3.3	267	3.5
Births and TOP[(c)]	88	4.6	79	4.1	74	3.8	69	3.6	310	4.0

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths. For births and terminations of pregnancy, the rate is per 10,000 live births and fetal deaths.

(c) Terminations of pregnancy <20 weeks gestation. TOP ≥ 20 weeks gestation or ≥ 400g birthweight are included as births.

Baby characteristics

Table 2.19.3: Births (gestation ≥20 weeks or birthweight ≥400g) with anorectal atresia/stenosis by baby characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 births
Sex			
Male	181	54.7	3.5
Female	145	43.8	3.0
Indeterminate	5	1.5	. .
Gestational age (weeks)			
20–36	115	34.7	14.9
37–41	213	64.4	2.3
42 and over	2	0.6	1.1
Not stated	1	0.3	. .
Birthweight (grams)			
Less than 2,500	119	36.0	17.7
2,500–4,499	208	62.8	2.3
4,500 and over	1	0.3	0.5
Not stated	3	0.9	. .

Maternal characteristics

Table 2.19.4: Number of women who gave birth to babies with anorectal atresia/stenosis, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Women who gave birth	92	3.7	77	3.1	82	3.3	80	3.2	331	3.3

(a) The rate is per 10,000 women who gave birth.

Table 2.19.5: Number of women who gave birth to babies with anorectal atresia/stenosis or who had a TOP where a fetus had this anomaly and the estimated rate, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Women who gave birth[(c)]	76	4.0	67	3.5	62	3.3	62	3.3	267	3.5
Women who gave birth or had a TOP[(d)]	88	4.6	79	4.1	74	3.9	69	3.7	310	4.1

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) The rate is per 10,000 women who gave birth.

(c) Includes women who had a termination of pregnancy at ≥ 20 weeks gestation.

(d) Women who had a termination of pregnancy at <20 weeks gestation.

Table 2.19.6: Births (gestation ≥ 20 weeks or birthweight ≥ 400g) with anorectal atresia/stenosis by maternal characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 women
Maternal age group			
Less than 20	22	6.6	4.5
20–24	54	16.3	3.5
25–29	117	35.3	3.7
30–34	88	26.6	2.9
35–39	41	12.4	2.9
40 and over	8	2.4	3.1
Not stated	1	0.3	. .
Indigenous status[1]			
Indigenous	19	5.8	6.5
Non-Indigenous	307	93.6	3.3
Not stated	2	0.6	. .
Remoteness Area			
Major cities	221	67.0	3.6
Regional	89	27.0	3.4
Remote	12	3.6	5.3
Not stated	8	2.4	. .
Country of birth			
Australia	258	77.9	4.3
New Zealand	4	1.2	2.8
United Kingdom	9	2.7	2.8
Europe	8	2.4	3.6
Middle East and North Africa	9	2.7	4.9
Asia	30	9.1	4.2
South and Central America and the Caribbean	1	0.3	2.0
Africa (excluding North Africa)	2	0.6	2.7
Other countries	4	1.2	4.4
Not stated	6	1.8	. .
Parity[2]			
Primiparous	106	42.6	4.0
Multiparous	140	56.2	3.6
Not stated	3	1.2	. .
Plurality			
Singleton	313	94.6	3.2
Multiple	18	5.4	11.4

(1) Excludes data from Tasmania

(2) Excludes data from New South Wales

Hirschsprung's disease

Definition: A condition characterised by partial or complete bowel obstruction resulting from absence of peristalsis in a segment of bowel due to an aganglionic section of the bowel.

ICD-9-BPA codes: 751.30–751.33

There were 1.3 cases of Hirschsprung's disease per 10,000 births reported between 1998 and 2001. There were no stillbirths or terminations of pregnancy reported for this condition during this period (Table 2.20.1).

Three-quarters of the babies born were male, 88.5% were born at term and the mean gestational age at birth was 38 completed weeks. The mean birthweight of the babies with Hirschsprung's disease was 3,380g and 92.4% weighed ≥2,500g (Table 2.20.3).

The mean maternal age of women who had a baby with Hirschsprung's disease was 28.6 years. More women living in major cities had an affected birth than the women living in remote areas (Table 2.20.6).

Reported rates of Hirschsprung's disease

Table 2.20.1: Number and rate of hirschsprung's disease by outcome, gestation ≥20 weeks or birthweight ≥400g, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Live births	33	1.3	31	1.2	31	1.2	36	1.4	131	1.3
Fetal deaths	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
All births	33	1.3	31	1.2	31	1.2	36	1.4	131	1.3

(a) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths.

Table 2.20.2: Number and the estimated rate of hirschsprung's disease by outcome, gestation ≥20 weeks or birthweight ≥400g and TOP <20 weeks, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Live births	27	1.4	23	1.2	26	1.4	29	1.5	105	1.4
Fetal deaths	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
All births	27	1.4	23	1.2	26	1.3	29	1.5	105	1.4
Births and TOP[(c)]	27	1.4	23	1.2	26	1.3	29	1.5	105	1.4

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths. For births and terminations of pregnancy, the rate is per 10,000 live births and fetal deaths.

(c) Terminations of pregnancy <20 weeks gestation. TOP ≥ 20 weeks gestation or ≥ 400g birthweight are included as births.

Baby characteristics

Table 2.20.3: Births (gestation ≥20 weeks or birthweight ≥400g) with hirschsprung's disease by baby characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 births
Sex			
Male	99	75.6	1.9
Female	32	24.4	0.7
Gestational age (weeks)			
20–36	15	11.5	1.9
37–41	116	88.5	1.3
Birthweight (grams)			
Less than 2,500	9	6.9	1.3
2,500–4,499	118	90.1	1.3
4,500 and over	3	2.3	1.6
Not stated	1	0.8	. .

Maternal characteristics

Table 2.20.4: Number of women who gave birth to babies with hirschsprung's disease, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Women who gave birth	33	1.3	31	1.2	31	1.2	36	1.5	131	1.3

(a) The rate is per 10,000 women who gave birth.

Table 2.20.5: Number of women who gave birth to babies with hirschsprung's disease or who had a TOP where a fetus had this anomaly and the estimated rate, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Women who gave birth[(c)]	27	1.4	23	1.2	26	1.4	29	1.5	105	1.4
Women who gave birth or had a TOP[(d)]	27	1.4	23	1.2	26	1.4	29	1.5	105	1.4

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) The rate is per 10,000 women who gave birth.

(c) Includes women who had a termination of pregnancy at ≥ 20 weeks gestation or ≥ 400g birthweight.

(d) Women who had a termination of pregnancy at <20 weeks gestation.

Table 2.20.6: Births (gestation ≥20 weeks or birthweight ≥400g) with hirschsprung's disease by maternal characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 women
Maternal age group			
Less than 20	8	6.1	1.7
20–24	25	19.1	1.6
25–29	40	30.5	1.3
30–34	35	26.7	1.1
35–39	20	15.3	1.4
40 and over	3	2.3	1.2
Indigenous status[1]			
Indigenous	3	2.3	1.0
Non-Indigenous	126	96.9	1.3
Not stated	1	0.8	. .
Remoteness Area			
Major cities	102	77.9	1.7
Regional	28	21.4	1.1
Remote	1	0.8	0.4
Country of birth			
Australia	100	76.3	1.7
New Zealand	4	3.1	2.8
United Kingdom	5	3.8	1.5
Europe	3	2.3	1.3
Middle East and North Africa	1	0.8	0.5
Asia	10	7.6	1.4
North America	1	0.8	2.0
Africa (excluding North Africa)	3	2.3	4.1
Not stated	4	3.1	. .
Parity[2]			
Primiparous	40	36.7	1.5
Multiparous	69	63.3	1.8
Plurality			
Singleton	129	98.5	1.3
Multiple	2	1.5	1.3

(1) Excludes data from Tasmania

(2) Excludes data from New South Wales

Hypospadias

Definition: a congenital anomaly characterised by the opening of the urethra on the ventral side of the penis, distally to the sulcus. Includes penile, scrotal, and perineal hypospadias. Exclude glanular or first-degree hypospadias and ambiguous genitalia (intersex or pseudohermaphroditism).

ICD-9-BPA codes: 752.60

ICD-10-AM codes: Q540–Q543, Q548, Q549 (ACT only)

Hypospadias is the most common congenital anomaly reported and this condition is limited to males. The severity of the condition was not reported to the national data collection. The overall rate of hypospadias was 25.9 per 10,000 births (Table 2.21.1). The rate of having an affected birth was 50.4 per 10,000 male births (Table 2.21.3).

The mean gestational age at birth was 38 completed weeks and 83.3% were born at term. The mean birthweight was 3,224g (Table 2.21.3).

The average age of women who gave birth to a baby with hypospadias was 29 years. This anomaly was more common in babies of non-Indigenous women and babies of women who had multiple births (Table 2.21.6).

Reported rates of hypospadias

Table 2.21.1: Number and rate of hypospadias by outcome, gestation ≥20 weeks or birthweight ≥400g, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Live births	675	27.0	629	25.0	678	26.9	622	25.0	2604	26.0
Fetal deaths	2	1.2	4	2.3	3	1.7	2	1.2	11	1.6
All births	677	26.9	633	24.9	681	26.9	624	24.9	2615	25.9

(a) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths.

Table 2.21.2: Number and the estimated rate of hypospadias by outcome, gestation ≥20 weeks or birthweight ≥400g and TOP <20 weeks, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Live births	558	29.2	548	28.4	538	28.0	522	27.6	2166	28.3
Fetal deaths	2	1.6	3	2.3	3	2.3	2	1.6	10	1.9
All births	560	29.1	551	28.4	541	27.9	524	27.5	2176	28.2
Births and TOP[(c)]	560	29.1	552	28.4	542	28.0	525	27.5	2179	28.3

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths. For births and terminations of pregnancy, the rate is per 10,000 live births and fetal deaths.

(c) Terminations of pregnancy <20 weeks gestation. TOP ≥ 20 weeks gestation or ≥ 400g birthweight are included as births.

Baby characteristics

Table 2.21.3: Births (gestation ≥20 weeks or birthweight ≥400g) by hypospadias by baby characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 births
Sex			
Male	2610	99.8	50.4
Female	0	0.0	. .
Indeterminate	5	0.2	. .
Gestational age (weeks)			
20–36	382	14.6	49.3
37–41	2178	83.3	23.8
42 and over	47	1.8	25.8
Not stated	8	0.3	. .
Birthweight (grams)			
Less than 2,500	381	14.6	56.7
2,500–4,499	2170	83.0	23.5
4,500 and over	55	2.1	29.5
Not stated	9	0.3	. .

Maternal characteristics

Table 2.21.4: Number of women who gave birth to babies with hypospadias, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Women who gave birth	676	27.3	629	25.2	681	27.3	620	25.2	2,606	26.2

(a) The rate is per 10,000 women who gave birth.

Table 2.21.5: Number of women who gave birth to babies with hypospadias or who had a TOP where a fetus had this anomaly and the estimated rate, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Women who gave birth[(c)]	559	29.5	548	28.7	541	28.4	520	27.7	2,168	28.6
Women who gave birth or had a TOP[(d)]	559	29.5	549	28.7	542	28.5	521	27.8	2,171	28.6

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) The rate is per 10,000 women who gave birth.

(c) Includes women who had a termination of pregnancy at ≥ 20 weeks gestation or ≥ 400g birthweight.

(d) Women who had a termination of pregnancy at <20 weeks gestation.

Table 2.21.6: Births (gestation ≥20 weeks or birthweight ≥400g) with hypospadias by maternal characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 women
Maternal age group			
Less than 20	122	4.7	25.2
20–24	412	15.8	26.3
25–29	844	32.4	27.0
30–34	794	30.5	25.8
35–39	362	13.9	25.5
40 and over	65	2.5	25.1
Not stated	7	0.3	. .
Indigenous status[(1)]			
Indigenous	59	2.3	20.3
Non-Indigenous	2521	97.3	26.8
Not stated	12	0.5	. .
Remoteness Area			
Major cities	1771	68.0	28.7
Regional	749	28.8	28.9
Remote	65	2.5	28.8
Not stated	19	0.7	. .
Country of birth			
Australia	2101	80.6	34.7
New Zealand	42	1.6	29.6
United Kingdom	102	3.9	31.4
Europe	71	2.7	31.8
Middle East and North Africa	86	3.3	46.3
Asia	101	3.9	14.2
North America	16	0.6	32.4
South and Central America and the Caribbean	4	0.2	7.9
Africa (excluding North Africa)	29	1.1	39.8
Other countries	8	0.3	8.7
Not stated	46	1.8	. .
Parity[(2)]			
Primiparous	912	45.4	34.5
Multiparous	1081	53.9	27.9
Not stated	14	0.7	. .
Plurality			
Singleton	2484	95.3	25.4
Multiple	122	4.7	77.3

(1) Excludes data from Tasmania

(2) Excludes data from New South Wales

Epispadias

Definition: a congenital anomaly characterised by the opening of the urethra on the dorsal surface of the penis. Not counted when part of exstrophy of the bladder.

ICD-9-BPA codes: 752.61

ICD-10-AM codes: Q64.0

The overall rate of epispadias was 0.5 per 10,000 births (Table 2.22.1). There were no stillbirths or terminations of pregnancy reported for this anomaly (Table 2.22.2).

Most babies were born at term (78.3%). The mean gestational age at birth was 38 weeks. The mean birthweight of babies born with epispadias was 3,081g (Table 2.22.3).

The mean maternal age of women who gave birth to a baby with epispadias was 30.6 years. Women aged 40 years or more had the highest rate of babies with epispadias. This condition was more common with multiple than singleton births (Table 2.22.6).

Reported rates of epispadias

Table 2.22.1: Number and rate of epispadias by outcome, gestation ≥20 weeks or birthweight ≥400g, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Live births	15	0.6	11	0.4	11	0.4	9	0.4	46	0.5
Fetal deaths	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
All births	15	0.6	11	0.4	11	0.4	9	0.4	46	0.5

(a) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths.

Table 2.22.2: Number and the estimated rate of epispadias by outcome, gestation ≥20 weeks or birthweight ≥400g and TOP <20 weeks, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Live births	13	0.7	8	0.4	10	0.5	7	0.4	38	0.5
Fetal deaths	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
All births	13	0.7	8	0.4	10	0.5	7	0.4	38	0.5
Births and TOP[(c)]	13	0.7	8	0.4	10	0.5	7	0.4	38	0.5

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths. For births and terminations of pregnancy, the rate is per 10,000 live births and fetal deaths.

(c) Terminations of pregnancy <20 weeks gestation. TOP ≥ 20 weeks gestation or ≥ 400g birthweight are included as births.

Baby characteristics

Table 2.22.3: Births (gestation ≥20 weeks or birthweight ≥400g) with epispadias by baby characteristics, Australia, 1998–2001

Characteristic	Number	Percent	Rate per 10,000 births
Sex			
Male	45	97.8	0.9
Female	0	0.0	0.0
Indeterminate	1	2.2	. .
Gestational age (weeks)			
20–36	10	21.7	1.3
37–41	36	78.3	0.4
Birthweight (grams)			
Less than 2,500	9	19.6	1.3
2,500–4,499	36	78.3	0.4
4,500 and over	1	2.2	0.5

Maternal characteristics

Table 2.22.4: Number of women who gave birth to babies with epispadias, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Women who gave birth	15	0.6	11	0.4	11	0.4	9	0.4	46	0.5

(a) The rate is per 10,000 women who gave birth.

Table 2.22.5: Number of women who gave birth to babies with epispadias or who had a TOP where a fetus had this anomaly and the estimated rate, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Women who gave birth[(c)]	13	0.7	8	0.4	10	0.5	7	0.4	38	0.5
Women who gave birth or had a TOP[(d)]	13	0.7	8	0.4	10	0.5	7	0.4	38	0.5

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) The rate is per 10,000 women who gave birth.

(c) Includes women who had a termination of pregnancy at ≥ 20 weeks gestation or ≥ 400g birthweight.

(d) Women who had a termination of pregnancy at <20 weeks gestation.

Table 2.22.6: Births (gestation ≥20 weeks or birthweight ≥400g) with epispadias by maternal characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 women
Maternal age group			
Less than 20	2	4.3	0.4
20–24	5	10.9	0.3
25–29	13	28.3	0.4
30–34	14	30.4	0.5
35–39	8	17.4	0.6
40 and over	4	8.7	1.5
Indigenous status[(1)]			
Indigenous	3	6.5	1.0
Non-Indigenous	43	93.5	0.5
Remoteness Area			
Major cities	36	78.3	0.6
Regional	8	17.4	0.3
Remote	1	2.2	0.4
Not stated	1	2.2	. .
Country of birth			
Australia	35	76.1	0.6
New Zealand	2	4.3	1.4
United Kingdom	2	4.3	0.6
Europe	1	2.2	0.4
Middle East and North Africa	2	4.3	1.1
Asia	2	4.3	0.3
North America	1	2.2	2.0
Not stated	1	2.2	. .
Parity[(2)]			
Primiparous	16	51.6	0.6
Multiparous	15	48.4	0.4
Plurality			
Singleton	44	95.7	0.5
Multiple	2	4.3	1.3

(1) Excludes data from Tasmania

(2) Excludes data from New South Wales

Renal agenesis and dysgenesis

Definition: A congenital anomaly characterised by complete absence of kidneys bilaterally or severely dysplastic kidneys.

ICD-9-BPA codes: 753.00–753.01

During the period 1998 to 2001, 5 cases of renal agenesis and dysgenesis per 10,000 births were reported. The data provided by four states that include termination of pregnancy before 20 weeks shows that 6.1 per 10,000 births were affected by this anomaly (Table 2.23.2).

Nearly two thirds of the births with this anomaly were males. The mean gestational age at birth was 34 completed weeks and 58.9% of those babies were born at term. The mean birthweight was 2,480g and more than half of the babies (58.3%) weighed ≥2,500g (Table 2.23.3).

Women living in remote areas and women who had multiple births had a higher rate of babies with this anomaly (Table 2.23.6).

Reported rates of renal agenesis and dysgenesis

Table 2.23.1: Number and rate of renal agenesis/dysgenesis by outcome, gestation ≥20 weeks or birthweight ≥400g, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Live births	113	4.5	100	4.0	91	3.6	112	4.5	416	4.1
Fetal deaths	21	12.2	22	12.6	25	14.1	20	11.6	88	12.6
All births	134	5.3	122	4.8	116	4.6	132	5.3	504	5.0

(a) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths.

Table 2.23.2: Number and the estimated rate of renal agenesis/dysgenesis by outcome, gestation ≥20 weeks or birthweight ≥400g and TOP <20 weeks, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Live births	94	4.9	80	4.1	72	3.7	80	4.2	326	4.3
Fetal deaths	16	12.4	20	15.4	21	15.9	17	13.3	74	14.3
All births	110	5.7	100	5.1	93	4.8	97	5.1	400	5.2
Births and TOP[(c)]	130	6.7	122	6.3	110	5.7	108	5.7	470	6.1

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths. For births and terminations of pregnancy, the rate is per 10,000 live births and fetal deaths.

(c) Terminations of pregnancy <20 weeks gestation. TOP ≥ 20 weeks gestation or ≥ 400g birthweight are included as births.

Baby characteristics

Table 2.23.3: Births (gestation ≥20 weeks or birthweight ≥400g) with renal agenesis/dysgenesis cases by baby characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 births
Sex			
Male	321	63.7	6.2
Female	174	34.5	3.5
Indeterminate	7	1.4	. .
Not stated	2	0.4	. .
Gestational age (weeks)			
20–36	202	40.1	26.1
37–41	291	57.7	3.2
42 and over	6	1.2	3.3
Not stated	5	1.0	. .
Birthweight (grams)			
Less than 2,500	203	40.3	30.2
2,500–4,499	289	57.3	3.1
4,500 and over	5	1.0	2.7
Not stated	7	1.4	. .

Maternal characteristics

Table 2.23.4: Number of women who gave birth to babies with renal agenesis/dysgenesis, Australia 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Women who gave birth	133	5.4	122	4.9	115	4.6	130	5.3	500	5.0

(a) The rate is per 10,000 women who gave birth.

Table 2.23.5: Number of women who gave birth to babies with renal agenesis/dysgenesis or who had a TOP where a fetus had this anomaly and the estimated rate, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Women who gave birth[(c)]	109	5.7	100	5.2	93	4.9	96	5.1	398	5.2
Women who gave birth or had a TOP[(d)]	129	6.8	122	6.4	110	5.8	107	5.7	468	6.2

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) The rate is per 10,000 women who gave birth.

(c) Includes women who had a termination of pregnancy at ≥ 20 weeks gestation or ≥ 400g birthweight.

(d) Women who had a termination of pregnancy at <20 weeks gestation.

Table 2.23.6: Births (gestation ≥20 weeks or birthweight ≥400g) with renal agenesis/dysgenesis by maternal characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 women
Maternal age group			
Less than 20	30	6.0	6.2
20–24	80	16.0	5.1
25–29	161	32.2	5.2
30–34	123	24.6	4.0
35–39	85	17.0	6.0
40 and over	16	3.2	6.2
Not stated	5	1.0	. .
Indigenous status[(1)]			
Indigenous	14	2.8	4.8
Non-Indigenous	472	95.4	5.0
Not stated	9	1.8	. .
Remoteness Area			
Major cities	342	68.4	5.6
Regional	130	26.0	5.0
Remote	18	3.6	8.0
Not stated	10	2.0	. .
Country of birth			
Australia	377	75.4	6.2
New Zealand	16	3.2	11.3
United Kingdom	16	3.2	4.9
Europe	8	1.6	3.6
Middle East and North Africa	21	4.2	11.3
Asia	25	5.0	3.5
North America	2	0.4	4.1
South and Central America and the Caribbean	7	1.4	13.9
Africa (excluding North Africa)	8	1.6	11.0
Other countries	3	0.6	3.3
Parity[(2)]			
Primiparous	143	37.6	5.4
Multiparous	230	60.5	5.9
Plurality			
Singleton	474	94.8	4.8
Multiple	26	5.2	16.5

(1) Excludes data from Tasmania

(2) Excludes data from New South Wales

Cystic kidney

Definition: A congenital anomaly characterised by multiple cysts in the kidney. Include infantile polycystic kidney, multicystic kidney, other forms of cystic kidney and unspecified cystic kidney. Exclude single kidney cyst.

ICD-9-BPA codes: 753.10–753.18

There were 4.7 per 10,000 births affected with cystic kidneys during 1998 to 2001 (Table 2.24.1). Data provided from the states that include termination of pregnancy before 20 weeks show that 5.8 per 10,000 births were affected by this anomaly (Table 2.24.2).

There were more males (60.1%) than females with cystic kidney. Two thirds of the births were at term and the mean gestational age at birth was 35 completed weeks. The average birthweight was 2,769g (Table 2.24.3).

The average age of women who had a baby with cystic kidney was 28.7 years. Women living in remote areas and women who had multiple births had higher rates of births with cystic kidney (Table 2.24.6).

Reported rates of Cystic kidney

Table 2.24.1: Number and rate of cystic kidney by outcome, gestation ≥20 weeks or birthweight ≥400g, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Live births	112	4.5	101	4.0	100	4.0	94	3.8	407	4.1
Fetal deaths	14	8.1	12	6.9	20	11.3	26	15.0	72	10.3
All births	126	5.0	113	4.5	120	4.7	120	4.8	479	4.7

(a) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths.

Table 2.24.2: Number and the estimated rate of cystic kidney by outcome, gestation ≥20 weeks or birthweight ≥400g and TOP <20 weeks, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Live births	84	4.4	79	4.1	79	4.1	78	4.1	320	4.2
Fetal deaths	12	9.3	11	8.5	17	12.8	22	17.1	62	11.9
All births	96	5.0	90	4.6	96	5.0	100	5.2	382	5.0
Births and TOP[(c)]	107	5.6	114	5.9	113	5.8	116	6.1	450	5.8

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths. For births and terminations of pregnancy, the rate is per 10,000 live births and fetal deaths.

(c) Terminations of pregnancy <20 weeks gestation. TOP ≥ 20 weeks gestation or ≥ 400g birthweight are included as births.

Baby characteristics

Table 2.24.3: Births (gestation ≥20 weeks or birthweight ≥400g) with cystic kidney by baby characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 births
Sex			
Male	288	60.1	5.6
Female	186	38.8	3.8
Indeterminate	1	0.2	. .
Not stated	4	0.8	. .
Gestational age (weeks)			
20–36	154	32.2	19.9
37–41	317	66.2	3.5
42 and over	4	0.8	2.2
Not stated	4	0.8	. .
Birthweight (grams)			
Less than 2,500	142	29.6	21.1
2,500–4,499	316	66.0	3.4
4,500 and over	14	2.9	7.5
Not stated	7	1.5	. .

Maternal characteristics

Table 2.24.4: Number of women who gave birth to babies with cystic kidney, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Women who gave birth	126	5.1	113	4.5	120	4.8	120	4.9	479	4.8

(a) The rate is per 10,000 women who gave birth.

Table 2.24.5: Number of women who gave birth to babies with cystic kidney or who had a TOP where a fetus had this anomaly and the estimated rate, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Women who gave birth[(c)]	96	5.1	90	4.7	96	5.0	100	5.3	382	5.0
Women who gave birth or had a TOP[(d)]	107	5.6	114	6.0	113	5.9	116	6.2	450	5.9

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) The rate is per 10,000 women who gave birth.

(c) Includes women who had a termination of pregnancy at ≥ 20 weeks gestation or ≥ 400g birthweight.

(d) Women who had a termination of pregnancy at <20 weeks gestation and <400g weight.

Table 2.24.6: Births (gestation ≥20 weeks or birthweight ≥400g) with cystic kidney by maternal characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 women
Maternal age group			
Less than 20	26	5.4	5.4
20–24	83	17.3	5.3
25–29	161	33.6	5.2
30–34	138	28.8	4.5
35–39	53	11.1	3.7
40 and over	11	2.3	4.3
Not stated	7	1.5	. .
Indigenous status[1]			
Indigenous	15	3.2	5.2
Non-Indigenous	449	94.5	4.8
Not stated	11	2.3	. .
Remoteness Area			
Major cities	326	68.3	5.3
Regional	126	26.4	4.9
Remote	15	3.1	6.6
Not stated	10	2.1	. .
Country of birth			
Australia	368	76.8	6.1
New Zealand	11	2.3	7.8
United Kingdom	19	4.0	5.9
Europe	12	2.5	5.4
Middle East and North Africa	12	2.5	6.5
Asia	30	6.3	4.2
North America	2	0.4	4.1
Africa (excluding North Africa)	6	1.3	8.2
Other countries	8	1.7	8.7
Not stated	11	2.3	. .
Parity[2]			
Primiparous	143	37.8	5.4
Multiparous	232	61.4	6.0
Not stated	3	0.8	. .
Plurality			
Singleton	458	95.6	4.7
Multiple	21	4.4	13.3

(1) Excludes data from Tasmania

(2) Excludes data from New South Wales

Bladder exstrophy

Definition: complex anomaly characterised by a defect in the closure of the lower abdominal wall and bladder. Bladder opens in the ventral wall of the abdomen between the umbilicus and the symphysis pubis. It is often associated with epispadias and structural anomalies of the pubic bones.

ICD-9-BPA codes: 753.5

Only a very small number of bladder exstrophy cases were reported during 1998 to 2001, the overall birth rate being 0.4 per 10,000 births. Very few stillbirths or terminations of pregnancy were reported (Table 2.25.1).

The mean gestational age at birth was 34 weeks. More than half (58.3%) of those affected babies were born at term. The mean birthweight was 2,453g (Table 2.25.3).

The mean maternal age of the women giving birth to a baby with bladder exstrophy was 28.6 years. There were no cases reported from babies born to Indigenous women. There were no reported cases from remote areas during the four years, 1998–2001. The anomaly rate was higher for multiple births than for singletons (Table 2.25.6).

Reported rates of Bladder exstrophy

Table 2.25.1: Number and rate of bladder exstrophy by outcome, gestation ≥20 weeks or birthweight ≥400g, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Live births	10	0.4	10	0.4	6	0.2	5	0.2	31	0.3
Fetal deaths	2	1.2	0	0.0	0	0.0	3	1.7	5	0.7
All births	12	0.5	10	0.4	6	0.2	8	0.3	36	0.4

(a) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths.

Table 2.25.2: Number and the estimated rate of bladder exstrophy by outcome, gestation ≥20 weeks or birthweight ≥400g and TOP <20 weeks, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Live births	9	0.5	7	0.4	6	0.3	5	0.3	27	0.4
Fetal deaths	1	0.8	0	0.0	0	0.0	2	1.6	3	0.6
All births	10	0.5	7	0.4	6	0.3	7	0.4	30	0.4
Births and TOP[(c)]	11	0.6	9	0.5	7	0.4	8	0.4	35	0.5

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths. For births and terminations of pregnancy, the rate is per 10,000 live births and fetal deaths.

(c) Terminations of pregnancy <20 weeks gestation. TOP ≥ 20 weeks gestation or ≥ 400g birthweight are included as births.

Baby characteristics

Table 2.25.3: Births (gestation ≥20 weeks or birthweight ≥400g) with bladder exstrophy by baby characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 births
Sex			
Male	22	61.1	0.4
Female	11	30.6	0.2
Indeterminate	3	8.3	. .
Gestational age (weeks)			
20–36	14	38.9	1.8
37–41	21	58.3	0.2
42 and over	1	2.8	0.5
Birthweight (grams)			
Less than 2,500	14	38.9	2.1
2,500–4,499	21	58.3	0.2
4,500 and over	1	2.8	0.5

Maternal characteristics

Table 2.25.4: Number of women who gave birth to babies with bladder exstrophy, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Women who gave birth	12	0.5	10	0.4	6	0.2	8	0.3	36	0.4

(a) The rate is per 10,000 women who gave birth.

Table 2.25.5: Number of women who gave birth to babies with bladder exstrophy or who had a TOP where a fetus had this anomaly and the estimated rate, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Women who gave birth[(c)]	10	0.5	7	0.4	6	0.3	7	0.4	30	0.4
Women who gave birth or had a TOP[(d)]	11	0.6	9	0.5	7	0.4	8	0.4	35	0.5

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) The rate is per 10,000 women who gave birth.

(c) Includes women who had a termination of pregnancy at ≥ 20 weeks gestation or ≥ 400g birthweight.

(d) Women who had a termination of pregnancy at <20 weeks gestation.

Table 2.25.6: Births (gestation ≥20 weeks or birthweight ≥400g) with bladder exstrophy by maternal characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 women
Maternal age group			
Less than 20	2	5.6	0.4
20–24	5	13.9	0.3
25–29	16	44.4	0.5
30–34	6	16.7	0.2
35–39	5	13.9	0.4
40 and over	2	5.6	0.8
Indigenous status[(1)]			
Indigenous	0	0.0	0.0
Non-Indigenous	36	100.0	0.4
Remoteness Area			
Major cities	24	66.7	0.4
Regional	10	27.8	0.4
Not stated	1	2.8	. .
Country of birth			
Australia	29	80.6	0.5
United Kingdom	3	8.3	0.9
Europe	1	2.8	0.4
Asia	2	5.6	0.3
Not stated	1	2.8	. .
Parity[(2)]			
Primiparous	8	34.8	0.3
Multiparous	15	65.2	0.4
Plurality			
Singleton	34	94.4	0.3
Multiple	2	5.6	1.3

(1) Excludes data from Tasmania

(2) Excludes data from New South Wales

Polydactyly

Definition: Extra digit(s) on the upper limb or the lower limb. It can affect the hand, the foot, or both.

ICD-9-BPA codes: 755.00–755.09

There were 8.9 per 10,000 births with polydactyly reported during 1998 to 2001.
More males than females had this anomaly (59.4%). The mean gestational age at birth was 37 completed weeks, with 81.9% of those babies born at term. A similar proportion (81.4%) had a birthweight between 2,500 to 4,499 g. The mean birthweight of babies with polydactyly was 3,138g (Table 2.26.3).

The average maternal age of those women who gave birth was 29 years. Women who had multiple births and Indigenous women had a higher rate of births with polydactyly (Table 2.26.6).

Reported rates of polydactyly

Table 2.26.1: Number and rate of polydactyly by outcome, gestation ≥20 weeks or birthweight ≥400g, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Live births	215	8.6	211	8.4	220	8.7	217	8.7	863	8.6
Fetal deaths	11	6.4	5	2.9	11	6.2	10	5.8	37	5.3
All births	226	9.0	216	8.5	231	9.1	227	9.1	900	8.9

(a) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths.

Table 2.26.2: Number and the estimated rate of polydactyly by outcome, gestation ≥20 weeks or birthweight ≥400g and TOP <20 weeks, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Live births	173	9.0	166	8.6	178	9.3	173	9.1	690	9.0
Fetal deaths	10	7.8	4	3.1	10	7.6	10	7.8	34	6.5
All births	183	9.5	170	8.8	188	9.7	183	9.6	724	9.4
Births and TOP[(c)]	191	9.9	179	9.2	198	10.2	194	10.2	762	9.9

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths. For births and terminations of pregnancy, the rate is per 10,000 live births and fetal deaths.

(c) Terminations of pregnancy <20 weeks gestation. TOP ≥ 20 weeks gestation or ≥ 400g birthweight are included as births.

Baby characteristics

Table 2.26.3: Births (gestation ≥20 weeks or birthweight ≥400g) with polydactyly by baby characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 births
Sex			
Male	535	59.4	10.3
Female	364	40.4	7.4
Indeterminate	1	0.1	. .
Gestational age (weeks)			
20–36	152	16.9	19.6
37–41	737	81.9	8.1
42 and over	10	1.1	5.5
Not stated	1	0.1	. .
Birthweight (grams)			
Less than 2,500	143	15.9	21.3
2,500–4,499	733	81.4	7.9
4,500 and over	23	2.6	12.3
Not stated	1	0.1	. .

Maternal characteristics

Table 2.26.4: Number of women who gave birth to babies with polydactyly, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Women who gave birth	226	9.1	216	8.6	231	9.3	227	9.2	900	9.1

(a) The rate is per 10,000 women who gave birth.

Table 2.26.5: Number of women who gave birth to babies with polydactyly or who had a TOP where a fetus had this anomaly and the estimated rate, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Women who gave birth[(c)]	183	9.6	170	8.9	188	9.9	183	9.8	724	9.5
Women who gave birth or had a TOP[(d)]	191	10.1	178	9.3	198	10.4	194	10.4	761	10.0

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) The rate is per 10,000 women who gave birth.

(c) Includes women who had a termination of pregnancy at ≥ 20 weeks gestation or ≥ 400g birthweight.

(d) Women who had a termination of pregnancy at <20 weeks gestation.

Table 2.26.6: Births (gestation ≥20 weeks or birthweight ≥400g) with polydactyly by maternal characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 women
Maternal age group			
Less than 20	48	5.3	9.9
20–24	167	18.6	10.7
25–29	258	28.7	8.3
30–34	257	28.6	8.3
35–39	133	14.8	9.4
40 and over	33	3.7	12.8
Not stated	4	0.4	. .
Indigenous status[1]			
Indigenous	35	3.9	12.0
Non-Indigenous	852	95.6	9.1
Not stated	4	0.4	. .
Remoteness Area			
Major cities	606	67.5	9.8
Regional	255	28.4	9.9
Remote	22	2.4	9.7
Not stated	15	1.7	. .
Country of birth			
Australia	635	70.6	10.5
New Zealand	31	3.4	21.9
United Kingdom	31	3.4	9.6
Europe	31	3.4	13.9
Middle East and North Africa	21	2.3	11.3
Asia	98	10.9	13.8
North America	0	0.0	0.0
South and Central America and the Caribbean	4	0.4	7.9
Africa (excluding North Africa)	16	1.8	22.0
Other countries	11	1.2	12.0
Not stated	22	2.4	. .
Parity[2]			
Primiparous	252	41.8	9.5
Multiparous	344	57.0	8.9
Not stated	7	1.2	. .
Plurality			
Singleton	873	97.0	8.9
Multiple	27	3.0	17.1

(1) Excludes data from Tasmania

(2) Excludes data from New South Wales

Limb reduction defects

Definition: A congenital anomaly characterised by total or partial absence or severe hypoplasia of skeletal structures of the limbs. Include femoral hypoplasia. Exclude mild hypoplasia with normal shape of skeletal parts, brachydactyly, finger or toe reduction directly associated with syndactyly, general skeletal dysplasia and sirenomelia.

ICD-9-BPA codes: 755.20–755.49

Limb reduction defects were seen in 5 per 10,000 births during this data collection period (Table 2.27.1). The rate for births and terminations of pregnancy from the four states providing TOP data was 6.6 per 10,000 births (Table 2.27.2).

Of the affected births 57% were males. The mean gestational age at birth was 35 completed weeks and two-thirds of them were born at term. The mean birthweight of those babies was 2,585g and 65.7% of the babies weighed ≥2,500g (Table 2.27.3).

The average maternal age was 28.1 years. Women aged less than 20 years had the highest rate of limb reduction defect births. Indigenous women had a higher rate of births with this anomaly compared to non-Indigenous women. Primiparous women and women who had multiple births, had higher birth rates with this condition (Table 2.27.6).

Reported rates of limb reduction defects

Table 2.27.1: Number and rate of limb reduction defects by outcome, gestation ≥20 weeks or birthweight ≥400g, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Live births	112	4.5	113	4.5	99	3.9	100	4.0	424	4.2
Fetal deaths	16	9.3	25	14.4	25	14.1	15	8.7	81	11.6
All births	128	5.1	138	5.4	124	4.9	115	4.6	505	5.0

(a) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths.

Table 2.27.2: Number and the estimated rate of limb reduction defects by outcome, gestation ≥20 weeks or birthweight ≥400g and TOP <20 weeks, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Live births	84	4.4	95	4.9	86	4.5	79	4.2	344	4.5
Fetal deaths	14	10.9	19	14.6	24	18.1	13	10.1	70	13.5
All births	98	5.1	114	5.9	110	5.7	92	4.8	414	5.4
Births and TOP[(c)]	124	6.4	150	7.7	127	6.6	110	5.8	511	6.6

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths. For births and terminations of pregnancy, the rate is per 10,000 live births and fetal deaths.

(c) Terminations of pregnancy <20 weeks gestation. TOP ≥ 20 weeks gestation or ≥ 400g birthweight are included as births.

Baby characteristics

Table 2.27.3: Births (gestation ≥20 weeks or birthweight ≥400g) with limb reduction defects by baby characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 births
Sex			
Male	288	57.0	5.6
Female	214	42.4	4.4
Indeterminate	2	0.4	. .
Not stated	1	0.2	. .
Gestational age (weeks)			
20–36	160	31.7	20.7
37–41	336	66.5	3.7
42 and over	8	1.6	4.4
Not stated	1	0.2	. .
Birthweight (grams)			
Less than 2,500	167	33.1	24.9
2,500–4,499	330	65.3	3.6
4,500 and over	2	0.4	1.1
Not stated	6	1.2	. .

Maternal characteristics

Table 2.27.4: Number of women who gave birth to babies with limb reduction defects, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Women who gave birth	128	5.2	138	5.5	124	5.0	114	4.6	504	5.1

(a) The rate is per 10,000 women who gave birth.

Table 2.27.5: Number of women who gave birth to babies with limb reduction defects or who had a TOP where a fetus had this anomaly and the estimated rate, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Women who gave birth[(c)]	98	5.2	114	6.0	110	5.8	91	4.9	413	5.4
Women who gave birth or had a TOP[(d)]	124	6.5	149	7.8	127	6.7	109	5.8	509	6.7

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) The rate is per 10,000 women who gave birth.

(c) Includes women who had a termination of pregnancy at ≥ 20 weeks gestation or ≥ 400g birthweight.

(d) Women who had a termination of pregnancy at <20 weeks gestation and <400g weight.

Table 2.27.6: Births (gestation ≥20 weeks or birthweight ≥400g) with limb reduction defects by maternal characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 women
Maternal age group			
Less than 20	44	8.7	9.1
20–24	97	19.2	6.2
25–29	146	29.0	4.7
30–34	134	26.6	4.3
35–39	67	13.3	4.7
40 and over	12	2.4	4.6
Not stated	4	0.8	. .
Indigenous status[1]			
Indigenous	22	4.4	7.6
Non-Indigenous	470	94.2	5.0
Not stated	7	1.4	. .
Remoteness Area			
Major cities	308	61.1	5.0
Regional	167	33.1	6.5
Remote	16	3.2	7.1
Not stated	13	2.6	. .
Country of birth			
Australia	404	80.2	6.7
New Zealand	13	2.6	9.2
United Kingdom	20	4.0	6.2
Europe	8	1.6	3.6
Middle East and North Africa	7	1.4	3.8
Asia	28	5.6	3.9
North America	1	0.2	2.0
South and Central America and the Caribbean	2	0.4	4.0
Africa (excluding North Africa)	5	1.0	6.9
Other countries	2	0.4	2.2
Not stated	14	2.8	. .
Parity[2]			
Primiparous	164	48.1	6.2
Multiparous	172	50.4	4.4
Not stated	5	1.5	. .
Plurality			
Singleton	482	95.6	4.9
Multiple	22	4.4	13.9

(1) Excludes data from Tasmania

(2) Excludes data from New South Wales

Diaphragmatic hernia

Definition: A congenital anomaly characterised by herniation into the thorax of abdominal contents through a defect of the diaphragm. Include total absence of the diaphragm. Exclude hiatus hernia, eventration and phrenic palsy.

ICD-9-BPA codes: 756.61

ICD-10-AM codes: Q79.0 (ACT only)

The diaphragmatic hernia was reported in 2.7 per 10,000 births during 1998 and 2001. There was a sudden increase in the rate in 1999 and it reverted back to the usual rate thereafter (Table 2.28.1 and Table 2.28.2). There were more males than females with this defect. The mean gestational age at birth was 35 completed weeks with two-thirds of the babies born at term. The mean birthweight for the babies born with this anomaly was 2,580g (Table 2.28.3).

The mean maternal age was 29.2 years. The rate of affected babies was higher among Indigenous women than non-Indigenous women. Women who had multiple births had a higher rate of babies with diaphragmatic hernia (Table 2.28.6).

Reported rates of Diaphragmatic hernia

Table 2.28.1: Number and rate of diaphragmatic hernia by outcome, gestation ≥20 weeks or birthweight ≥400g, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Live births	59	2.4	78	3.1	48	1.9	57	2.3	242	2.4
Fetal deaths	8	4.6	8	4.6	6	3.4	11	6.4	33	4.7
All births	67	2.7	86	3.4	54	2.1	68	2.7	275	2.7

(a) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths.

Table 2.28.2: Number and the estimated rate of diaphragmatic hernia by outcome, gestation ≥20 weeks or birthweight ≥400g and TOP <20 weeks, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Live births	40	2.1	65	3.4	36	1.9	40	2.1	181	2.4
Fetal deaths	6	4.7	6	4.6	6	4.5	8	6.2	26	5.0
All births	46	2.4	71	3.7	42	2.2	48	2.5	207	2.7
Births and TOP[(c)]	54	2.8	87	4.5	53	2.7	54	2.8	248	3.2

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths. For births and terminations of pregnancy, the rate is per 10,000 live births and fetal deaths.

(c) Terminations of pregnancy <20 weeks gestation. TOP ≥ 20 weeks gestation or ≥ 400g birthweight are included as births.

Baby characteristics

Table 2.28.3: Births (gestation ≥20 weeks or birthweight ≥400g) with diaphragmatic hernia by baby characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 births
Sex			
Male	167	60.7	3.2
Female	105	38.2	2.1
Indeterminate	3	1.1	. .
Gestational age (weeks)			
20–36	95	34.5	12.3
37–41	176	64.0	1.9
42 and over	4	1.5	2.2
Birthweight (grams)			
Less than 2,500	96	34.9	14.3
2,500–4,499	173	62.9	1.9
4,500 and over	1	0.4	0.5
Not stated	5	1.8	. .

Maternal characteristics

Table 2.28.4: Number of women who gave birth to babies with diaphragmatic hernia, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Women who gave birth	67	2.7	86	3.4	54	2.2	68	2.8	275	2.8

(a) The rate is per 10,000 women who gave birth.

Table 2.28.5: Number of women who gave birth to babies with diaphragmatic hernia or who had an induced abortion where a fetus had this anomaly and the estimated rate, selected states[(a)], 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Women who gave birth[(c)]	46	2.4	71	3.7	42	2.2	48	2.6	207	2.7
Women who gave birth or had a TOP[(d)]	54	2.8	87	4.6	53	2.8	54	2.9	248	3.3

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) The rate is per 10,000 women who gave birth.

(c) Includes women who had a termination of pregnancy at ≥ 20 weeks gestation or ≥ 400g birthweight.

(d) Women who had a termination of pregnancy at <20 weeks gestation.

Table 2.28.6: Births (gestation ≥20 weeks or birthweight ≥400g) with diaphragmatic hernias by maternal characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 women
Maternal age group			
Less than 20	17	6.2	3.5
20–24	39	14.2	2.5
25–29	87	31.6	2.8
30–34	78	28.4	2.5
35–39	44	16.0	3.1
40 and over	8	2.9	3.1
Not stated	2	0.7	. .
Indigenous status[(1)]			
Indigenous	13	4.7	4.5
Non-Indigenous	258	94.2	2.7
Not stated	3	1.1	. .
Remoteness Area			
Major cities	186	67.6	3.0
Regional	77	28.0	3.0
Remote	7	2.5	3.1
Not stated	5	1.8	. .
Country of birth			
Australia	201	73.1	3.3
New Zealand	8	2.9	5.6
United Kingdom	13	4.7	4.0
Europe	9	3.3	4.0
Middle East and North Africa	6	2.2	3.2
Asia	23	8.4	3.2
North America	1	0.4	2.0
South and Central America and the Caribbean	4	1.5	7.9
Africa (excluding North Africa)	2	0.7	2.7
Other countries	2	0.7	2.2
Not stated	6	2.2	. .
Parity[(2)]			
Primiparous	87	46.0	3.3
Multiparous	100	52.9	2.6
Not stated	2	1.1	. .
Plurality			
Singleton	268	97.5	2.7
Multiple	7	2.5	4.4

(1) Excludes data from Tasmania

(2) Excludes data from New South Wales

Exomphalos

Definition: A congenital anomaly characterised by herniation of abdominal contents through the umbilical insertion and covered by a membrane which may or may not be intact. Exclude gastroschisis, hypoplasia of abdominal muscles and skin covered umbilical hernia.

ICD-9-BPA codes: 756.70

ICD-10-AM codes: Q79.2 (ACT only)

There were 1.5 exomphalos cases per 10,000 births in 1998 to 2001(Table 2.29.1). There was a twofold increase in this rate when the TOP before 20 weeks data were added. However, the rate of exomphalos in births and TOP from the four states has decreased from 4 per 10,000 births in 1998 to 2.9 per 10,000 births in 2001 (Table 2.29.2).

There were more males than females with this anomaly. The average gestational age was 32 completed weeks, and more than half (54.5%) of affected babies were born preterm. The mean birthweight was 2,169g, and half of the babies weighed less than 2,500g (Table 2.29.3).

The average age of women who had a baby with exomphalos was 29.5 years. Women aged 40 years or more and women who had multiple births had a higher rate of births with exomphalos (Table 2.29.6).

Reported rates of exomphalos

Table 2.29.1: Number and rate of exomphalos by outcome, gestation ≥20 weeks or birthweight ≥400g, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Live births	28	1.1	28	1.1	27	1.1	27	1.1	110	1.1
Fetal deaths	10	5.8	12	6.9	10	5.6	14	8.1	46	6.6
All births	38	1.5	40	1.6	37	1.5	41	1.6	156	1.5

(a) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths.

Table 2.29.2: Number and the estimated rate of exomphalos by outcome, gestation ≥20 weeks or birthweight ≥400g and TOP <20 weeks, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Live births	25	1.3	21	1.1	20	1.0	21	1.1	87	1.1
Fetal deaths	10	7.8	10	7.7	9	6.8	14	10.9	43	8.3
All births	35	1.8	31	1.6	29	1.5	35	1.8	130	1.7
Births and TOP[(c)]	78	4.0	48	2.5	55	2.8	56	2.9	237	3.1

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths. For births and terminations of pregnancy, the rate is per 10,000 live births and fetal deaths.

(c) Terminations of pregnancy <20 weeks gestation. TOP ≥ 20 weeks gestation or ≥ 400g birthweight are included as births.

Baby characteristics

Table 2.29.3: Births (gestation ≥20 weeks or birthweight ≥400g) with exomphalos by baby characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 births
Sex			
Male	92	59.0	1.8
Female	62	39.7	1.3
Indeterminate	2	1.3	. .
Gestational age (weeks)			
20–36	85	54.5	11.0
37–41	69	44.2	0.8
42 and over	2	1.3	1.1
Birthweight (grams)			
Less than 2,500	79	50.6	11.8
2,500–4,499	74	47.4	0.8
4,500 and over	2	1.3	1.1
Not stated	1	0.6	. .

Maternal characteristics

Table 2.29.4: Number of women who gave birth to babies with exomphalos, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Women who gave birth	38	1.5	40	1.6	37	1.5	40	1.6	155	1.6

(a) The rate is per 10,000 women who gave birth.

Table 2.29.5: Number of women who gave birth to babies with exomphalos or who had a TOP where a fetus had this anomaly and the estimated rate, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Women who gave birth[(c)]	35	1.8	31	1.6	29	1.5	34	1.8	129	1.7
Women who gave birth or had a TOP[(d)]	78	4.1	48	2.5	55	2.9	55	2.9	236	3.1

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) The rate is per 10,000 women who gave birth.

(c) Includes women who had a termination of pregnancy at ≥ 20 weeks gestation or ≥ 400g birthweight.

(d) Women who had a termination of pregnancy at <20 weeks gestation.

Table 2.29.6: Births (gestation ≥20 weeks or birthweight ≥400g) with exomphalos by maternal characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 women
Maternal age group			
Less than 20	9	5.8	1.9
20–24	21	13.5	1.3
25–29	50	32.3	1.6
30–34	38	24.5	1.2
35–39	25	16.1	1.8
40 and over	10	6.5	3.9
Not stated	2	1.3	. .
Indigenous status[1]			
Indigenous	5	3.2	1.7
Non-Indigenous	147	95.5	1.6
Not stated	2	1.3	. .
Remoteness Area			
Major cities	85	55.6	1.4
Regional	60	39.2	2.3
Remote	6	3.9	2.7
Not stated	2	1.3	. .
Country of birth			
Australia	123	79.4	2.0
New Zealand	3	1.9	2.1
United Kingdom	3	1.9	0.9
Europe	4	2.6	1.8
Middle East and North Africa	2	1.3	1.1
Asia	9	5.8	1.3
North America	1	0.6	2.0
South and Central America and the Caribbean	1	0.6	2.0
Other countries	4	2.6	4.4
Not stated	5	3.2	. .
Parity[2]			
Primiparous	53	51.0	2.0
Multiparous	48	46.2	1.2
Not stated	3	2.9	. .
Plurality			
Singleton	148	95.5	1.5
Multiple	7	4.5	4.4

(1) Excludes data from Tasmania

(2) Excludes data from New South Wales

Gastroschisis

Definition: A congenital anomaly characterised by visceral herniation through a right side abdominal wall defect to an intact umbilical cord and not covered by a membrane. Exclude hypoplasia of abdominal muscles, skin covered umbilical hernia and exomphalos.

ICD-9-BPA codes: 756.71

ICD-10-AM codes: Q79.3 (ACT only)

There were 2.6 cases of gastroschisis per 10,000 births reported during 1998 to 2001. Very few stillbirths or terminations of pregnancy were reported during this period (Table 2.30.1 and Table 2.30.2).

There were more males than females with this abnormality. The mean gestational age was 35 completed weeks, with more than half of those babies born preterm. 54.7% had a birthweight less than 2,500g with the average of affected cases being 2,321g (Table 2.30.3).

The mean maternal age of women with affected babies was 23.3 years. This condition was more common when the maternal age was <25 years. Indigenous women, primiparous women and women who had multiple births had higher rates of gastroschisis births (Table 2.30.6).

Reported rates of gastroschisis

Table 2.30.1: Number and rate of gastroschisis by outcome, gestation ≥20 weeks or birthweight ≥400g, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Live births	52	2.1	59	2.3	63	2.5	70	2.8	244	2.4
Fetal deaths	6	3.5	7	4.0	4	2.3	6	3.5	23	3.3
All births	58	2.3	66	2.6	67	2.6	76	3.0	267	2.6

(a) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths.

Table 2.30.2: Number and the estimated rate of gastroschisis by outcome, gestation ≥20 weeks or birthweight ≥400g and TOP <20 weeks, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Live births	38	2.0	42	2.2	42	2.2	46	2.4	168	2.2
Fetal deaths	6	4.7	5	3.8	3	2.3	6	4.7	20	3.9
All births	44	2.3	47	2.4	45	2.3	52	2.7	188	2.4
Births and TOP[(c)]	47	2.4	49	2.5	49	2.5	55	2.9	200	2.6

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths. For births and terminations of pregnancy, the rate is per 10,000 live births and fetal deaths.

(c) Terminations of pregnancy <20 weeks gestation. TOP ≥ 20 weeks gestation or ≥ 400g birthweight are included as births.

Baby characteristics

Table 2.30.3: Births (gestation ≥20 weeks or birthweight ≥400g) with gatroschisis by baby characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 births
Sex			
Male	156	58.4	3.0
Female	109	40.8	2.2
Indeterminate	2	0.7	. .
Gestational age (weeks)			
20–36	146	54.7	18.9
37–41	120	44.9	1.3
42 and over	1	0.4	0.5
Birthweight (grams)			
Less than 2,500	151	56.6	22.5
2,500–4,499	116	43.4	1.3

Maternal characteristics

Table 2.30.4: Number of women who gave birth to babies with gatroschisis, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Women who gave birth	58	2.3	66	2.6	67	2.7	76	3.1	267	2.7

(a) The rate is per 10,000 women who gave birth.

Table 2.30.5: Number of women who gave birth to babies with gatroschisis or who had a TOP where a fetus had this anomaly and the estimated rate, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Women who gave birth[(c)]	44	2.3	47	2.5	45	2.4	52	2.8	188	2.5
Women who gave birth or had a TOP[(d)]	47	2.5	49	2.6	49	2.6	55	2.9	200	2.6

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) The rate is per 10,000 women who gave birth.

(c) Includes women who had a termination of pregnancy at ≥ 20 weeks gestation or ≥ 400g birthweight.

(d) Women who had a termination of pregnancy at <20 weeks gestation.

Table 2.30.6: Births (gestation ≥20 weeks or birthweight ≥400g) with gastroschisis by maternal characteristics Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 women
Maternal age group			
Less than 20	66	24.7	13.6
20–24	109	40.8	7.0
25–29	57	21.3	1.8
30–34	29	10.9	0.9
35–39	6	2.2	0.4
Indigenous status[(1)]			
Indigenous	11	4.2	3.8
Non-Indigenous	249	94.0	2.6
Not stated	5	1.9	. .
Remoteness Area			
Major cities	157	59.2	2.5
Regional	101	38.1	3.9
Remote	6	2.3	2.7
Not stated	1	0.4	. .
Country of birth			
Australia	234	87.6	3.9
New Zealand	3	1.1	2.1
United Kingdom	5	1.9	1.5
Europe	2	0.7	0.9
Middle East and North Africa	3	1.1	1.6
Asia	9	3.4	1.3
North America	1	0.4	2.0
South and Central America and the Caribbean	1	0.4	2.0
Africa (excluding North Africa)	1	0.4	1.4
Other countries	3	1.1	3.3
Not stated	5	1.9	. .
Parity[(2)]			
Primiparous	129	66.2	4.9
Multiparous	65	33.3	1.7
Not stated	1	0.5	. .
Plurality			
Singleton	258	96.6	2.6
Multiple	9	3.4	5.7

Trisomy 13

Definition: A congenital chromosomal malformation syndrome associated with extra chromosome 13 material. Include translocation and mosaic trisomy 13.

ICD-9-BPA codes: 758.10–758.19

The overall rate of trisomy 13 births was 0.8 per 10,000 births (Table 2.31.1). The rate of births and terminations of pregnancy for trisomy 13 from the four states providing TOP data was 2.5 per 10,000 births (Table 2.31.2) indicating a higher rate of terminations.

The proportion of males to female for trisomy 13 cases was roughly equal. The mean gestational age at birth was 29 weeks and most (73.4%) were born preterm. The mean birthweight was 1,462g, with 75.9% of affected babies having a birthweight less than 2,500g (Table 2.31.3).

The mean maternal age was 30.4 years. The rate of trisomy 13 births appeared to increase with advancing maternal age. Indigenous women had a higher rate of births with this condition compared to non-Indigenous women (Table 2.31.6).

Reported rates of trisomy 13

Table 2.31.1: Number and rate of trisomy 13 by outcome, gestation ≥20 weeks or birthweight ≥400g, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Live births	13	0.5	10	0.4	12	0.5	6	0.2	41	0.4
Fetal deaths	10	5.8	8	4.6	10	5.6	10	5.8	38	5.4
All births	23	0.9	18	0.7	22	0.9	16	0.6	79	0.8

(a) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths.

Table 2.31.2: Number and the estimated rate of trisomy 13 by outcome, gestation ≥20 weeks or birthweight ≥400g and TOP <20 weeks, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Live births	9	0.5	10	0.5	10	0.5	4	0.2	33	0.4
Fetal deaths	8	6.2	6	4.6	8	6.0	9	7.0	31	6.0
All births	17	0.9	16	0.8	18	0.9	13	0.7	64	0.8
Births and TOP[(c)]	40	2.1	47	2.4	57	2.9	46	2.4	190	2.5

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths. For births and terminations of pregnancy, the rate is per 10,000 live births and fetal deaths.

(c) Terminations of pregnancy <20 weeks gestation. TOP ≥ 20 weeks gestation or ≥ 400g birthweight are included as births.

Baby characteristics

Table 2.31.3: Births (gestation ≥20 weeks or birthweight ≥400g) with trisomy 13 by baby characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 births
Sex			
Male	41	51.9	0.8
Female	37	46.8	0.8
Indeterminate	1	1.3	. .
Gestational age (weeks)			
20–36	58	73.4	7.5
37–41	21	26.6	0.2
Birthweight (grams)			
Less than 2,500	60	75.9	8.9
2,500–4,499	18	22.8	0.2
Not stated	1	1.3	. .

Maternal characteristics

Table 2.31.4: Number of women who gave birth to babies with trisomy 13, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Women who gave birth	23	0.9	18	0.7	22	0.9	16	0.6	79	0.8

(a) The rate is per 10,000 women who gave birth.

Table 2.31.5: Number of women who gave birth to babies with trisomy 13 or who had a TOP where a fetus had this anomaly and the estimated rate, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Women who gave birth[(c)]	17	0.9	16	0.8	18	0.9	13	0.7	64	0.8
Women who gave birth or had a TOP[(d)]	40	2.1	47	2.5	57	3.0	46	2.5	190	2.5

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) The rate is per 10,000 women who gave birth.

(c) Includes women who had a termination of pregnancy at ≥ 20 weeks gestation or ≥ 400g birthweight.

(d) Women who had a termination of pregnancy at <20 weeks gestation.

Table 2.31.6: Births (gestation ≥20 weeks or birthweight ≥400g) with trisomy 13 by maternal characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 women
Maternal age group			
Less than 20	3	3.8	0.6
20–24	12	15.2	0.8
25–29	19	24.1	0.6
30–34	21	26.6	0.7
35–39	19	24.1	1.3
40 and over	5	6.3	1.9
Indigenous status[(1)]			
Indigenous	4	5.1	1.4
Non-Indigenous	73	92.4	0.8
Not stated	2	2.5	. .
Remoteness Area			
Major cities	53	67.9	0.9
Regional	23	29.5	0.9
Remote	2	2.6	0.9
Country of birth			
Australia	57	72.2	0.9
New Zealand	6	7.6	4.2
United Kingdom	2	2.5	0.6
Europe	4	5.1	1.8
Asia	4	5.1	0.6
Africa (excluding North Africa)	1	1.3	1.4
Other countries	1	1.3	1.1
Not stated	4	5.1	. .
Parity[(2)]			
Primiparous	12	25.0	0.5
Multiparous	36	75.0	0.9
Plurality			
Singleton	78	98.7	0.8
Multiple	1	1.3	0.6

(1) Excludes data from Tasmania

(2) Excludes data from New South Wales

Trisomy 18

Definition: A congenital chromosomal malformation syndrome associated with extra chromosome 18. Include translocation and mosaic trisomy 18.

ICD-9-BPA codes: 758.20–758.29

The reported rate of trisomy 18 at birth was 2.0 per 10,000 births (Table 2.32.1). The rate of births and terminations of pregnancy at <20 weeks from the states providing data, was three times higher than the rate at birth (6.3 per 10,000 births, Table 2.32.2).

There were slightly more females (53.2%) than males with trisomy 18. The mean gestational age was 31 weeks and nearly two-thirds were born preterm. The mean birthweight of affected babies was 1,282g. 89.3% weighed less than 2,500g (Table 2.32.3).

The mean maternal age was 32.7 years. The rate of having a birth with this anomaly has increased with advancing maternal age. The rate of trisomy 18 births was higher for those women having multiple births (Table 2.32.6).

Reported rates of trisomy 18

Table 2.32.1: Number and rate of trisomy 18 by outcome, gestation ≥20 weeks or birthweight ≥400g, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Live births	27	1.1	30	1.2	15	0.6	23	0.9	95	0.9
Fetal deaths	32	18.5	30	17.2	27	15.2	21	12.2	110	15.8
All births	59	2.3	60	2.4	42	1.7	44	1.8	205	2.0

(a) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths.

Table 2.32.2: Number and the estimated rate of trisomy 18 by outcome, gestation ≥20 weeks or birthweight ≥400g and TOP <20 weeks, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Live births	19	1.0	22	1.1	14	0.7	15	0.8	70	0.9
Fetal deaths	30	23.3	27	20.8	25	18.9	18	14.0	100	19.3
All births	49	2.5	49	2.5	39	2.0	33	1.7	170	2.2
Births and TOP[(c)]	126	6.5	129	6.6	118	6.1	115	6.0	488	6.3

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths. For births and terminations of pregnancy, the rate is per 10,000 live births and fetal deaths.

(c) Terminations of pregnancy <20 weeks gestation. TOP ≥ 20 weeks gestation or ≥ 400g birthweight are included as births.

Baby characteristics

Table 2.32.3: Births (gestation ≥20 weeks or birthweight ≥400g) with trisomy 18 by baby characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 births
Sex			
Male	95	46.3	1.8
Female	109	53.2	2.2
Not stated	1	0.5	. .
Gestational age (weeks)			
20–36	128	62.4	16.5
37–41	68	33.2	0.7
42 and over	6	2.9	3.3
Not stated	3	1.5	. .
Birthweight (grams)			
Less than 2,500	183	89.3	27.2
2,500–4,499	14	6.8	0.2
4,500 and over	1	0.5	0.5
Not stated	7	3.4	. .

Maternal characteristics

Table 2.32.4: Number of women who gave birth to babies with trisomy 18, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Women who gave birth	59	2.4	60	2.4	42	1.7	43	1.7	204	2.1

(a) The rate is per 10,000 women who gave birth.

Table 2.32.5: Number of women who gave birth to babies with trisomy 18 or who had a TOP where a fetus had this anomaly and the estimated rate, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Women who gave birth[(c)]	49	2.6	49	2.6	39	2.0	32	1.7	169	2.2
Women who gave birth or had a TOP[(d)]	126	6.6	129	6.7	117	6.1	112	6.0	484	6.4

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) The rate is per 10,000 women who gave birth.

(c) Includes women who had a termination of pregnancy at ≥ 20 weeks gestation or ≥ 400g birthweight.

(d) Women who had a termination of pregnancy at <20 weeks gestation.

Table 2.32.6: Births (gestation ≥20 weeks or birthweight ≥400g) with trisomy 18 by maternal characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 women
Maternal age group			
Less than 20	8	3.9	1.7
20–24	15	7.4	1.0
25–29	42	20.6	1.3
30–34	52	25.5	1.7
35–39	48	23.5	3.4
40 and over	35	17.2	13.5
Not stated	4	2.0	. .
Indigenous status[(1)]			
Indigenous	7	3.5	2.4
Non-Indigenous	185	92.0	2.0
Not stated	9	4.5	. .
Remoteness Area			
Major cities	131	64.9	2.1
Regional	53	26.2	2.0
Remote	7	3.5	3.1
Not stated	11	5.4	. .
Country of birth			
Australia	126	61.8	2.1
New Zealand	1	0.5	0.7
United Kingdom	11	5.4	3.4
Europe	10	4.9	4.5
Middle East and North Africa	5	2.5	2.7
Asia	28	13.7	3.9
North America	1	0.5	2.0
South and Central America and the Caribbean	1	0.5	2.0
Africa (excluding North Africa)	2	1.0	2.7
Other countries	6	2.9	6.5
Not stated	13	6.4	. .
Parity[(2)]			
Primiparous	45	33.6	1.7
Multiparous	86	64.2	2.2
Not stated	3	2.2	. .
Plurality			
Singleton	195	95.6	2.0
Multiple	9	4.4	5.7

(1) Excludes data from Tasmania

(2) Excludes data from New South Wales

Trisomy 21

Definition: A congenital chromosomal malformation syndrome characterised by a well known pattern of minor and major anomalies and associated with excess chromosomal 21 material. Include trisomy mosaicism and translocations of chromosome 21.

ICD-9-BPA codes: 758.00–758.09

The overall rate of trisomy 21 across 1998 to 2001 was 11.5 per 10,000 births. The data provided by four states that include termination of pregnancy before 20 weeks shows that 23.3 per 10,000 births were affected by this anomaly.

There were more males (55.7%) than females with trisomy 21. The average gestational age at birth was 35 weeks, and about two thirds were born at term. The average birthweight was 2,624g and two thirds had a birthweight ≥2,500g (Table 2.33.3).

Mean maternal age of women who had a baby with trisomy 21 was 32.5 years. The rate of having a baby with trisomy 21 increased with the advancing maternal age. The Trisomy 21 birth rate was greater for women who had multiple births compared to singleton births (Table 2.33.6).

Reported rates of Down syndrome

Table 2.33.1: Number and rate of trisomy 21 by outcome, gestation ≥20 weeks or birthweight ≥400g, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Live births	244	9.8	240	9.5	291	11.6	246	9.9	1,021	10.2
Fetal deaths	35	20.3	46	26.4	30	16.9	34	19.7	145	20.8
All births	279	11.1	286	11.3	321	12.7	280	11.2	1,166	11.5

(a) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths.

Table 2.33.2: Number and the estimated rate of trisomy 21 by outcome, gestation ≥20 weeks or birthweight ≥400g and TOP <20 weeks, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
Outcome	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Live births	187	9.8	181	9.4	220	11.4	181	9.6	769	10.0
Fetal deaths	33	25.7	38	29.3	26	19.7	25	19.5	122	23.5
All births	220	11.4	219	11.3	246	12.7	206	10.8	891	11.6
Births and TOP[(c)]	407	21.1	454	23.4	490	25.3	442	23.2	1,793	23.3

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) For all births, the rate is per 10,000 live births and fetal deaths. For live births, the rate is per 10,000 live births and for fetal deaths, the rate is per 1,000 fetal deaths. For births and terminations of pregnancy, the rate is per 10,000 live births and fetal deaths.

(c) Terminations of pregnancy <20 weeks gestation. TOP ≥ 20 weeks gestation or ≥ 400g birthweight are included as births.

Baby characteristics

Table 2.33.3: Births (gestation ≥20 weeks or birthweight ≥400g) with trisomy 21 by baby characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 births
Sex			
Male	650	55.7	12.5
Female	511	43.8	10.4
Indeterminate	1	0.1	. .
Not stated	4	0.3	. .
Gestational age (weeks)			
20–36	387	33.2	50.0
37–41	734	63.0	8.0
42 and over	7	0.6	3.8
Not stated	38	3.3	. .
Birthweight (grams)			
Less than 2,500	362	31.0	53.9
2,500–4,499	742	63.6	8.0
4,500 and over	6	0.5	3.2
Not stated	56	4.8	. .

Maternal characteristics

Table 2.33.4: Number of women who gave birth to babies with trisomy 21, Australia, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]	Number	Rate[(a)]
Women who gave birth	279	11.3	286	11.4	320	12.8	279	11.3	1,164	11.7

(a) The rate is per 10,000 women who gave birth.

Table 2.33.5: Number of women who gave birth to babies with trisomy 21 or who had a TOP where a fetus had this anomaly and the estimated rate, available for four states[(a)] only, 1998–2001

	1998		1999		2000		2001		1998–2001	
	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]	Number	Rate[(b)]
Women who gave birth[(c)]	220	11.6	219	11.5	246	12.9	206	11.0	891	11.7
Women who gave birth or had a TOP[(d)]	405	21.3	453	23.7	485	25.5	440	23.5	1,783	23.5

(a) Includes data for New South Wales, Victoria, Western Australia and South Australia.

(b) The rate is per 10,000 women who gave birth.

(c) Includes women who had a termination of pregnancy at ≥ 20 weeks gestation or ≥ 400g birthweight.

(d) Women who had a termination of pregnancy at <20 weeks gestation and <400g weight.

Table 2.33.6: Births (gestation ≥20 weeks or birthweight ≥400g) with trisomy 21 by maternal characteristics, Australia, 1998–2001

Characteristic	Number	Per cent	Rate per 10,000 women
Maternal age group			
Less than 20	25	2.1	5.2
20–24	89	7.6	5.7
25–29	229	19.7	7.3
30–34	345	29.6	11.2
35–39	278	23.9	19.6
40 and over	141	12.1	54.5
Not stated	57	4.9	. .
Indigenous status[1]			
Indigenous	30	2.6	10.3
Non-Indigenous	1049	90.8	11.2
Not stated	76	6.6	. .
Remoteness Area			
Major cities	720	61.9	11.7
Regional	378	32.5	14.6
Remote	31	2.7	13.7
Not stated	35	3.0	. .
Country of birth			
Australia	813	69.8	13.4
New Zealand	25	2.1	17.6
United Kingdom	44	3.8	13.6
Europe	32	2.7	14.3
Middle East and North Africa	29	2.5	15.6
Asia	103	8.8	14.5
North America	8	0.7	16.2
South and Central America and the Caribbean	2	0.2	4.0
Africa (excluding North Africa)	7	0.6	9.6
Other countries	14	1.2	15.3
Parity[2]			
Primiparous	260	34.1	9.8
Multiparous	492	64.5	12.7
Not stated	11	1.4	. .
Plurality			
Singleton	1127	96.8	11.5
Multiple	36	3.1	22.8

(1) Excludes data from Tasmania

(2) Excludes data from New South Wales

4 Australian Paediatric Surveillance Unit — selected statistics

The APSU is a national research resource that facilitates the collection of national data on uncommon childhood conditions through active, monthly surveillance with reporting from child health specialists (Elliott E, et al. 2005). Child health specialists notify cases to the APSU and case notification is followed up by a questionnaire providing detailed demographic and clinical data. Cases are classified according to a predefined case definition after exclusion of errors and duplicate reports.

Data on selected conditions characterised by birth defects are presented below. Additional information about the Australian Paediatric Surveillance Unit is presented in Chapter 1 and available at <http://www.apsu.org.au>

In this chapter, data are presented for Prader-Willi syndrome, fetal alcohol syndrome, CHARGE association, congenital arthrogryposis (AMC), congenital varicella and congenital rubella syndrome. Prader-Willi syndrome, CHARGE association and some types of AMC are genetic disorders. Fetal alcohol syndrome and congenital rubella, and congenital varicella are preventable birth defects.

Prader-Willi syndrome

Prader-Willi syndrome (PWS) is a genetic disorder in which an abnormality of chromosome 15 is seen in virtually all cases. The usual mechanism of genetic abnormality is either uniparental disomy or DNA deletion. Newborns with PWS are typically severely hypotonic and feed poorly. Development is delayed and children often have hypogonadism, typical facial features and excessive sleepiness. Behavioural and obesity-related problems develop in childhood (Smith et al. 2003).

Study period

January 1998 to December 2000.

Case definition

Clinicians were asked to report any children aged <15 years with newly diagnosed PWS. Diagnosis could be made clinically or following genetic investigation (karyotype, FISH test or methylation test).

Selected statistics

There were 42 confirmed cases of PWS in the period 1998–2000. The age at diagnosis ranged from birth to 11 years. Males accounted for 63.4% (26) of cases. Of the 42 cases, 71.4% were aged <1 year at diagnosis and 60.0% of these cases were male. Their median age at diagnosis was one month.

Uniparental disomy was the mechanism of genetic abnormality in 29.0% of cases and DNA deletion in 54.0%. In 17.0% the genetic mechanism was unknown.

Table 3.1.1 Selected statistics from the study on Prader-Willi syndrome, 1998–2000

Selected statistics			Number	Per cent
Surveillance				
	Response rate [a]		94	74.6
Cases				
	Confirmed Prader-Willi syndrome [b]		42	
Estimated incidence rate [c]			0.4	
Characteristics				
	Males		26	63.4
	Females		15	36.6
	Age at diagnosis (median; range)	1 month;	birth to 11 years	. .
	Infants aged <1 year at diagnosis		30	71.4
			Males	18
			Females	12

Source: Elliott et al. 2001

(a) Response rate is the proportion of notifications for which questionnaires containing clinical data were returned.

(b) Final number of cases are those that met predetermined case definition criteria and excludes any reporting errors and duplicate reports

(c) Rate per 100,000 population aged <15 years per annum

Prader-Willi syndrome was confirmed in 0.4 per 100,000 children under 15 years of age during the period 1998 to 2000.

Data were available for 41 out of 42 confirmed cases. DNA testing was performed for all 41 cases. DNA tests used include chromosomal testing, methylation testing and fluorescence in situ hybridisation (FISH). This study recommends that the methylation test be the 'gold-standard' in PWS diagnostic testing. This test should be performed first, followed by FISH test.

Excessive sleepiness is the only PWS phenotype that was shown to be significantly associated with a particular genetic mechanism in this study.

Table 3.1.2 Clinical features of Prader Willi syndrome by genetic mechanism

Genetic mechanism	UPD	Deletion	unknown
Major clinical features			
Severe neonatal hypotonia	8 (67%)	16 (73%)	7 (100%)
Unusual facial features	9 (75%)	20 (91%)	5 (71%)
Never breastfed	8 (67%)	10 (46%)	4 (57%)
Hypogonadal	9 (75%)	17 (77%)	2 (29%)
Minor clinical features			
Decreased fetal movements	6 (50%)	11 (50%)	3 (43%)
Breech/transverse presentation	4 (33%)	5 (23%)	3 (43%)
Polyhydramnios	1 (8%)	2 (9%)	0
Excessive sleepiness	9 (75%)	7 (32%)	4 (57%)
Eye abnormality	1 (8%)	6 (27%)	4 (57%)
Behavioural problems	3 (25%)	1 (5%)	0
Food related problems	2 (17%)	2 (9%)	1 (14%)
Small hands	4 (33%)	8 (36%)	4 (57%)
Small feet	6 (50%)	7 (31%)	4 (57%)
Other features			
Caesarean section delivery	8 (67%)	8 (36%)	2 (29%)
Resuscitation at birth	5 (42%)	4 (18%)	2 (29%)
Hypopigmentation	3 (25%)	7 (31%)	2 (29%)

Source: Elliott et al. 2001

Fetal alcohol syndrome

Fetal alcohol syndrome (FAS) is at the most severe end of the spectrum of birth defects caused by maternal alcohol consumption in early pregnancy. FAS is characterised by minor cranio-facial abnormalities, prenatal and/or postnatal growth deficiency and evidence of damage to or dysfunction of the central nervous system (Elliott et al. 2005).

Study period

January 2001 to December 2004.

Case definition

In this study, clinicians were asked to report any child aged <15 years with newly diagnosed FAS according to the following definitions:

Fetal alcohol syndrome – alcohol exposure confirmed

Evidence of prenatal alcohol exposure AND all characteristic cranio-facial abnormalities AND prenatal or postnatal growth deficiency AND structural abnormalities or dysfunction to the central nervous system.

Partial Fetal alcohol syndrome – alcohol exposure confirmed

Evidence of prenatal alcohol exposure AND all characteristic cranio-facial abnormalities AND prenatal or postnatal growth deficiency OR some characteristic cranio-facial abnormalities AND structural abnormalities or dysfunction to the central nervous system.

Suspected Fetal alcohol syndrome – alcohol exposure not confirmed

All characteristic cranio-facial abnormalities AND prenatal or postnatal growth deficiency AND structural abnormalities or dysfunction to the CNS.

Selected statistics

There were 92 confirmed cases of FAS in the period 2001–2004 and an increase in case reports by year over the study period. Of the 92 confirmed cases, 25 (27.2%) were classified as FAS, 65 (70.6%) as partial FAS and 2 (2.2%) suspected FAS. The median age of children at diagnosis was 3.3 years and this ranged from 1 day to 11 years. Males accounted for 53.0% of cases and 65.0% of cases were identified as Indigenous.

Over a third (35.0%) of the children were preterm (<37 weeks gestation) and 65.0% were of low birthweight (<2.5kg). Most (94.0%) had 'high risk' exposure to alcohol in utero and 78.0% were exposed to one or more additional drugs. Of the 92 cases, 56.0% had growth deficiency, 53.0% microcephaly, 84.0% CNS dysfunction, 24.0% additional birth defects, 5.0% sensorineural deafness and 4.0% visual impairment. Most (83.7%) had behavioural, cognitive or emotional problems. In addition to features required for diagnosis, 24.0% of children had features consistent with 'alcohol-related birth defects.' These included musculoskeletal abnormalities (cervical rib, proximal placement of thumbs, pectus carinatum, thoracic kyphosis, talipes, spina

bifida occulta, persistently patent fontanelles) in seven children; cardiovascular abnormalities (VSD, ASD, PDA, PS, PFA, vascular ring, haemangioma) in eight; renal abnormalities (horseshoe kidney, vesicoureteric reflux, renal dysplasia) in two; cleft palate in two; and microphthalmia in one child. Half of the children (51.0%) had a sibling with FAS suggesting missed opportunities for prevention. Only 40.0% lived with a biological parent. Children with FAS used required support services including remedial education, specialist therapy and multiple health services.

Table 3.2: Selected statistics from the study on fetal alcohol syndrome, 2001–2004

Selected statistics		Number	Per cent
Surveillance			
	Response rate [a]	159	94.1
Cases [b]			
	FAS	25	32.9
	Partial FAS	49	64.5
	Suspected FAS	2	2.6
Total		76	100
Characteristics			
	Males	39	51.3
	Females	37	48.7
	Indigenous	46	60.5
	Non-Indigenous	30	39.5
	Median age at diagnosis (years)	2.8	. .
	Age at diagnosis (range)	Birth to 12 years	. .
Estimated incidence rate [c]			
Australia (Indigenous + non-Indigenous)		0.58	
Indigenous children		7.4	
Non-Indigenous children		0.22	

Source: Elliott et al. 2006 and Elliott et al. 2005.

(a) Response rate is the proportion of notifications for which questionnaires containing clinical data were returned.

(b) Final number of cases are those that met pre-determined case definition criteria and excludes any reporting errors and duplicate reports

(c) Rate per 100,000 population aged <15 years per annum

Fetal alcohol syndrome was diagnosed in 0.58 per 100,000 children under 15 years of age during the period 2001 to 2004. Indigenous children had a higher rate of fetal alcohol syndrome compared to non-Indigenous children. The rate for Indigenous children was 7.4 per 100,000 children and for non-Indigenous children 0.22 per 100,000 children.

CHARGE association

CHARGE association (coloboma-heart disease-atresia of choanae, retarded mental development and growth, genital hypoplasia, ear abnormalities-deafness) is a non-random pattern of congenital abnormalities including coloboma, choanal atresia and cardiac, renal, genital and gastrointestinal abnormalities. Diagnosis is hindered by inconsistencies in diagnostic criteria and may be delayed because major features of the disorder may not be readily identifiable, particularly in young children. CHARGE association shares many features with other genetic and birth defect syndromes (Elliott et al. 2005).

Study period

January 2000 to December 2002.

Case definition

In this study, clinicians were asked to report any newly diagnosed cases in children <16 years of age. Revised consensus criteria were used (Blake et al 1998). Children with three or more major criteria alone or children with one or more major criteria with at least two minor criteria were included.

Clinical features - major and minor criteria used for this study

Major criteria:

- coloboma – iris, retina or choroid
- choanal atresia – unilateral or bilateral; membranous or bony; stenosis or atresia
- characteristic ear abnormalities – lop or cup shaped external year; middle ear (ossicular) malformations, chronic serous otitis, mixed deafness, cochlear defects
- cranial nerve dysfunction – anosmia; facial palsy; sensori neural deafness; vestibular problems and swallowing difficulties.

Minor criteria:

- genital hypoplasia – males: micropenis, cryptorchidism; Females: hypoplastic labia; males and females: delayed and incomplete pubertal development
- developmental delay – delayed motor milestones, hypotonia, mental retardation
- cardiovascular malformations – all types: especially conotruncal defects and atrioventricular canal defects
- orofacial clefts – cleft lip and/or palate
- tracheo-oesophageal fistula – tracheo-oesophageal defects of all types
- distinctive face – unilateral facial weakness/asymmetry, broad forehead, flat malar region and small mid face, thin lips

Selected statistics

There were 23 cases of CHARGE association identified in the period 2000–2002 (Table 3.3). Males accounted for 47.8% of cases. The median age at diagnosis was 12 days (range 1 to 75 days). Cases were reported in all jurisdictions except the Northern Territory. The number of cases ranged from one each in Victoria, South Australia, Tasmania and the Australian Capital Territory to eight each in New South Wales and Queensland. Three cases were reported in Western Australia. All children presented with a complex of abnormalities that fulfilled the CHARGE diagnostic criteria. The most common of the major criteria were characteristic ear abnormalities (95.7%) and coloboma (91.3%). The most common of the minor criteria was cardiovascular malformations occurring in 82.6% of cases.

The estimated incidence of CHARGE association in Australia was 2.8 per 100,000 live births during the period 2000 to 2002.

Table 3.3: Selected statistics from the study on CHARGE association, 2000–2002

Selected statistics		Number	Per cent
Surveillance			
	Response rate [a]	57	93.4
Cases[b]			
	Confirmed CHARGE association	23	. .
Characteristics			
	Males	11	47.8
	Females	12	52.2
	Median age at diagnosis	12 days	. .
	Age at diagnosis (range)	1–75 days	. .
Criteria for diagnosis (major)			
	Coloboma	21	91.3
	Choanal atresia	14	60.9
	Characteristic ear abnormalities	22	95.7
	Cranial nerve dysfunction	17	73.9
Criteria for diagnosis (minor)			
	Genital hypoplasia	12	52.2
	Developmental delay[c]	10	43.5
	Cardiovascular malformations	19	82.6
	Orofacial clefts	6	6.8
	Tracheo-oesophageal fistula	3	13
	Distinctive face	12	52.2

(a) Response rate is the proportion of notifications for which questionnaires containing clinical data were returned.

(b) Final number of cases are those that met predetermined case definition criteria and excludes any reporting errors and duplicate reports

(c) Does not include data on the six children who died.

Congenital arthrogryposis (AMC) (syndrome)

Arthrogryposis multiplex congenita (AMC) describes the presence of multiple joint contractures at birth. A joint contracture is fixed high resistance to passive straightening of the joint. The term AMC is used to cover a diverse range of conditions in which multiple joint contractures occur.

Study period

1 January 1996 to 31 December 1998.

Case definition

This study included any child born during the study period with two or more non-progressive joint contractures present since birth.

Selected statistics

Forty-seven cases of AMC were identified during the study period (Table 3.4). Of these children, 43 (91.5%) had bilateral involvement of two or more major joint sites (elbow, wrist, hand, hip, knee or ankle) signifying severe disease. Nine children had contractures of the fingers in addition to major joint involvement. Forty children (84.1%) had a variety of birth defects in addition to the joint contractures (Table 3.4). Ten of these had an identifiable syndrome and two had a chromosomal abnormality – one trisomy 18 and the other had an unbalanced reciprocal translocation between chromosomes 6 and 9.

Prenatal factors associated with AMC were identified in 12 children and included: oligohydramnios; breech presentation; polyhydramnios with prolonged rupture of membranes; intrauterine growth retardation; bicornuate uterus; maternal myasthenia gravis; small placenta and twin pregnancy.

Eleven children had a contributory family history including: consanguinity (4); sibling affected by AMC (2); maternal history of myasthenia gravis, multiple epiphysial dysplasia and significant intellectual disability of unknown aetiology; father and paternal uncle required splints in infancy (possibly a milder form of the disorder); and one father had a balanced reciprocal translocation between chromosomes 6 and 9.

The incidence of AMC was 6.2 per 100,000 liveborn infants during this study period. This could be an underestimate because stillbirths and termination of pregnancies due to AMC were not counted for these calculations.

Table 3.4 Selected statistics from the study on arthrogryposis multiplex congenita, 1996–1998

Selected statistics		Number	Percent
Surveillance			
	Response rate [a]	96	95
Cases			
	Confirmed AMC [b]	47	. .
Estimated incidence rate [c]		6.2	. .
Characteristics			
	Males	23	47.8
	Females	24	52.2
Associated clinical problem			
	No additional clinical problem	7	14.9
	Additional clinical problem	40	84.1
	Identifiable syndrome	10	25
	Chromosomal abnormality	2	5
	Other problems	28	70
Prenatal factor			
	Yes	12	25.5
	No	35	74.5
Contributory family history			
	Yes	11	25.7
	No	36	74.3

Source: Elliott et al.1999.

(a) Response rate is the proportion of notifications for which questionnaires containing clinical data were returned.

(b) Final number of cases are those that met predetermined case definition criteria and excludes any reporting errors and duplicate reports

(c) Rate per 100,000 live births per annum

Congenital rubella

Rubella infection in women in the first trimester of pregnancy can result in congenital rubella syndrome (CRS) with birth defects in the fetus, including deafness, cataracts, growth retardation mental handicap and cardiac abnormalities (Forrest et al. 2003).

Study period

January 1993 to December 2007 (ongoing).

Case definition

Clinicians were asked to report any child aged <16 years of age with newly diagnosed congenital rubella. The case definition for this study was:

Any child who, in the opinion of the notifying paediatrician, has definite or suspected congenital rubella, with or without defects, based on history, clinical or laboratory findings.

Reported birth defects

Most (~80%) of the children with CRS had significant, often multiple defects including: deafness, cataracts, retinopathy, keratitis, microcephaly, intracerebral calcification, cardiovascular malformations such as patent ductus arteriosus and pulmonary artery stenosis, pneumonitis, intrauterine growth retardation, failure to thrive and developmental delay (Elliott et al 2001).

Selected statistics

There were 106 notifications of congenital rubella in the period 1993–2006 with 57 cases: 50 (87.7%) confirmed and 7 (12.3%) probable.

For 29 (58% of confirmed cases), maternal infection occurred in Australia and for 8 cases (22.2%), maternal infection occurred overseas. In addition, there were seven cases of children diagnosed with CRS in Australia who were born overseas in Kenya, Nauru, Fiji and Indonesia (Forrest et al. 2003). For cases where maternal infection occurred in Australia, 16 (55.2%) were born in New South Wales, 8 (27.6%) were born in Queensland and the remainder were born in Victoria (3) and the Australian Capital Territory (2).

Data from the APSU study informed a change to the immunisation schedule in 1998 to include a second dose at age five prior to school entry for both boys and girls using the combined measles, mumps rubella (MMR) vaccine. Fewer cases of CRS were reported from 1998 to 2000. However, since 2001 six infants were born in Australia with CRS – four to overseas-born women and two due to missed school immunisation in Australian-born women (Forrest et al. 2003; Elliott et al. 2005).

Table 3.5 Selected statistics from the study on Congenital Rubella, 1993–2006

Selected statistics		Number	Percent
Surveillance			
	Response rate [(a)]	102	96.2
Cases [(b)]			
	Confirmed congenital rubella	50	87.7
	Probable congenital rubella	7	12.3
Total		57	100
Estimated incidence rate [(c)] (confirmed cases only)		0.1	
Infection acquired in Australia (confirmed cases)		29	
	Mother born overseas	9	77.8

Source: Elliott et al.2006 and Forrest et al. 2003.

(a) Response rate is the proportion of notifications for which questionnaires containing clinical data were returned.

(b) Final number of cases are those that met predetermined case definition criteria and excludes any reporting errors and duplicate reports

(c) Rate per 100,000 population aged <15 years per annum

There have been no reports of congenital rubella in 2005 and 2006. However, with increased migration into Australia of women from countries with poor immunisation programs, continued surveillance is essential (Zurynski et al. 2005).

Congenital varicella

Congenital varicella may lead to spontaneous abortion or premature delivery and may also cause embryopathy. Hypoplasia of one limb with cicatricial skin lesions affecting that limb in a dermatomal distribution, intrauterine growth retardation, neurological abnormalities (including: microcephaly, hydrocephalus and cerebellar hypoplasia, motor and sensory deficits and sphincter dysfunction), eye lesions (including microphthalmia, cataracts, Horner's syndrome, chorioretinitis, retinal scars, nystagmus and optic atrophy) and occasional gastrointestinal (hepatic failure) and genito-urinary abnormalities are typical features of the congenital varicella syndrome.

Study period

March 1995 to December 1997.

Case definition

The case definition for this study was any stillborn, newborn infant or child up to the age of two years who, in the opinion of the notifying paediatrician, has definite or suspected congenital varicella, with or without defects, based on history, clinical and laboratory findings.

Summary statistics

There were seven confirmed cases of congenital varicella (including one termination due to congenital varicella) between 1995 and 1997. Maternal infection at between 8 and 26 weeks gestation resulted in severe malformations. The malformations included the following: limb, skeletal, heart, central nervous system and gastrointestinal defects, skin scars, eye damage including chorioretinitis and blindness. Birth defects were reported even when maternal infection occured late in pregnancy. The incidence of congenital varicella was 0.8 per 100,000 live births per annum. (Elliott E, et al. 1998; Forrest et al. 2000)

Table 3.6 Clinical features of congenital varicella cases 1995–1997

Case	Year of birth	Timing of maternal varicella in pregnancy (weeks)	Defects and other clinical features
1	1995	13	No defects; disseminated zoster at 5 months - recovered
2	1995	18	No defects; zoster at 15 months
3	1995	26	Skin scars; chorioretinitis
4	1996	12	Skin scars; blindness; zoster at 10 weeks of age
5	1997	8	Severe limb, skeletal, heart and central and peripheral nervous system defects. Died at 10 days of age
6	1997	11	Skin scars hydrocephalus; pregnancy terminated at 21 weeks after ultrasound showed hydrocephalus
7	1997	16	Skin scars, neurological and gastrointestinal defects

Source: Forrest et al.2000; Elliott et al.1998

Current study

The Australian National Varicella Vaccination Program commenced in November 2005. APSU reactivated surveillance of congenital varicella in Australia in 2006, enabling this new study to provide a unique opportunity to compare current rates of congenital and neonatal infection and the sources of infection in Australia to rates reported in 1995–1997 (Peadon et al. 2006).

5 Future directions

National Minimum Data Set for congenital anomalies

The AIHW National Perinatal Statistics Unit (NPSU) has received Australian Health Minister's Advisory Council (AHMAC) funding through the Statistical Information Management Committee (SIMC) to work to develop a National Minimum Data Set (NMDS) for congenital anomalies.

A NMDS is a core set of data elements agreed to by the SIMC and endorsed by the National Health Information Management Principal Committee (NHIMPC) for mandatory collection and reporting at a national level. An NMDS depends on a national agreement to collect uniform data and to supply it as part of a national collection (HDSC 2006). The standards make data collection activities more efficient by reducing duplication of effort by standardising core data items; more effective by ensuring that information to be collected is relevant and appropriate to its purpose; and more comparable and consistent for reporting purposes. An NMDS includes agreement on specified data elements (discrete items of information or variables) and supporting data element concepts as well as the application of those data elements and the statistical units for collection. Definitions of all data elements that are included in National Minimum Data Sets are included in the AIHW's online metadata registry, 'METeOR'.

The development of a NMDS for congenital anomalies will involve developing and agreeing on:

- a definition for congenital anomalies
- the scope of the NMDS, including the conditions to be included, the period of notification and the data elements to be included
- the data element definitions
- a classification for congenital anomalies
- a time-frame for implementation
- integration with the perinatal NMDS

Some jurisdictions use ICD-9-BPA to code congenital anomalies and others use ICD-10-AM. The jurisdictions using ICD-9-BPA have not moved to ICD-10-AM because it lacks specificity for some congenital anomalies. Following the review of the National Congenital Malformations and Birth Defects Data Collection, the National Congenital Anomalies Steering Committee (NCASC) agreed that a single classification should be developed for use in Australia and that this should be based on ICD-10-AM. NPSU is working with the National Centre for Classification in Health (NCCH) to develop a classification for congenital anomalies based on the seventh edition of ICD-10-AM. Consultation with state and territory experts on coding congenital anomalies will be undertaken.

NPSU is also working with NCCH and state and territory experts to develop maps from ICD-9-BPA to ICD-10-AM so that data can be presented in a single classification in national reports and to enable time series analyses to be undertaken using a single classification.

Committees

The NMDS for Congenital anomalies will be developed in consultation with several committees convened by the NPSU – NCASC, State and Territory Implementation Committee for Congenital Anomalies (STICCA) (See Appendix D) and when relevant, National Perinatal Data Development Committee (NPDDC). Following endorsement by NCASC and STICCA, submissions will be made to the Health Data Standards Committee (HDSC) and SIMC.

Proposed timeframe

The proposed timeframe for the development of the NMDS for congenital anomalies is by December 2008, with implementation from 1 July 2009. The time-frame for the development of the classification for congenital anomalies is by early 2009.

Clinical definitions

For some of the conditions included in this report, which were defined by the International Clearinghouse for Birth Defects Surveillance and Research (ICBDSR), the definitions were thought to be inadequate for the Australian context by a working group of the NCASC. The Australian Paediatric Surveillance Unit (APSU) is currently undertaking a project, in consultation with NPSU, on developing nationally standardised clinical definitions for selected congenital anomalies that are adequate for the Australian context and will provide more accurate information about the prevalence of these conditions.

Congenital anomalies in Australia 2002–2003

A report on congenital anomalies for 2002 and 2003 is currently being prepared and will be published later in 2007.

National report on neural tube defects

Food Standards Australia and New Zealand (FSANZ) has approved mandatory fortification of food with folic acid to reduce the prevalence of neural tube defects (NTD). An important part of assessing the impact of fortification is the availability of baseline data to compare rates of NTD pre- and post-fortification. A national report on NTD using currently available data is being prepared. It will contain data on births and induced abortions with NTD occurring in 1998–2003. This report will be published in December 2007.

Newborn screening

Newborn screening has been undertaken in Australia since the late 1960's. It identifies a range of metabolic disorders in newborn babies for which morbidity, mortality and associated disabilities can be reduced by early intervention (HGSA-RACP 2004). A blood sample is taken from a newborn baby between 48 and 72 hours after birth by pricking the baby's heel and collecting blood onto absorbent paper (a Guthrie card). Since the 1998 the scope of testing has been greatly expanded by using tandem mass spectrometry, which is now universal throughout Australia.

Prenatal screening and diagnosis

The aim of prenatal screening and diagnostic testing is the early detection of congenital anomalies. This enables strategies for prevention and treatment to be developed early on, thus reducing the impact of the disease.

Prenatal screening programs differ from diagnostic testing in that screening programs are conducted at a population level and, in the context of congenital anomalies, are used to identify pregnant women that may be at risk of having a baby with a congenital anomaly.

Screening programs may be targeted at 'high risk' individuals, for example pregnant women aged 37 years and over. Diagnostic testing is conducted at the individual level to diagnose or confirm a birth anomaly. Indications for diagnostic testing include advanced maternal age (37 years and over), abnormal screening results, history of a congenital anomaly and exposure to a teratogen (Muggli & Halliday 2003).

Prenatal screening tests include: full blood examinations; first and second trimester maternal serum screening (this is a biochemical test which is usually done in the second trimester, but is being increasingly performed in the first trimester to coincide with the measurement of nuchal translucency conducted by ultrasound); ultrasound including nuchal translucency (this is performed at 8–12 weeks to determine gestational age and at 10–14 weeks to determine the nuchal translucency which can indicate a risk of chromosomal abnormality); ultrasound performed in the second trimester can be used to screen for structural congenital anomalies.

Prenatal diagnostic tests include: amniocentesis, which involves the collection of amniotic fluid from the amniotic cavity in the uterus. The cells from the amniotic fluid are grown in culture and chromosomal, biochemical and molecular biological analyses are undertaken to detect various birth anomalies and certain genetic diseases. This test is performed at 14–18 weeks of pregnancy; chorionic billus sampling, which involves the collection of tissue from the villi of the chorion (part of the placenta) and testing for congenital anomalies and genetic diseases. This test is performed at 10–12 weeks of pregnancy, so can be performed earlier than amniocentesis.

National reporting of data on newborn screening and prenatal screening/diagnosis

The review of the National Congenital Malformations and Birth Defects Data Collection (Birch et al. 2004) recommended that NPSU develop collaborations with organisations collecting data on newborn screening and prenatal screening/diagnosis and that data on newborn screening and prenatal screening/diagnosis be included in the revised national report on congenital anomalies.

The NPSU has received AHMAC funding through SIMC to prepare two options papers, on how to proceed with national data collection and reporting on newborn screening and prenatal screening/diagnosis. The purpose of this project is to conduct a scoping study on the need for and feasibility of developing a Data Set Specification (DSS), which is a set of metadata that are not mandated for collection but are recommended as best practice, or NMDS for newborn screening and for prenatal screening/diagnosis.

The NPSU will consult with the State and Territory Working Group of the AHMAC Advisory Group, state and territory health authorities, HGSA/RACP and other stakeholders for the options papers.

Appendix A: Denominator data

Table A: Confinements, births and deaths, all states and territories except Northern Territory

	1998	1999	2000	2001	1998-2001
Women who gave birth	247840	249813	249439	246342	993434
Fetal deaths	1728	1742	1775	1728	6973
Live births	249952	252075	251792	248828	1002647
Not stated	118	17	4	0	139
All births	**251680**	**253817**	**253567**	**250556**	**1009620**

Table B: Confinements, births and deaths: NSW, Vic, SA and WA only

	1998	1999	2000	2001	1998-2001
Women who gave birth	189749	191149	190425	187408	247840
Fetal deaths	1286	1299	1323	1283	1728
Live births	191402	192952	192259	189367	249952
Not stated	118	17	4	0	118
All births	**192688**	**194251**	**193582**	**190650**	**251680**

Table C: Indigenous status, Australia, 1998–2001

Aboriginal or Torres Strait Islander	29073
Other	940761
Not stated	147
Total	969981

Note: Excludes Northern Territory and Tasmania data

Table D: Remoteness Area of usual residence, Australia, 1998–2001

Major cities of Australia	566744
Total regional	215960
otal remote	17425
Not stated	320
Total	800513

Note: Excludes Northern Territory and Queensland data

Table E: Parity, 1998–2001

Primiparous	264115
Multiparous	387536
Not stated	29
Total	651680

Note: Excludes Northern Territory and New South Wales data

Appendix B: State and territory reports on congenital anomalies

New South Wales

Centre for Epidemiology and Research 2004. New South Wales Mothers and Babies 2003. NSW Public Health Bulletin 15: S-5. Sydney: NSW Department of Health.

Victoria

Riley M & Halliday J 2006. Birth defects in Victoria 2003–2004. Victorian Perinatal Data Collection Unit. Public Health. Melbourne: Department of Human Services.

Muggli E & Halliday J 2006. Report on prenatal diagnostic testing in Victoria 2005. Public Health Genetics, Murdoch Children's Research Institute.

Queensland

Queensland Health 2007. Perinatal statistics Queensland 2005. Brisbane: Queensland Health.

South Australia

Hann E, Chan A, Scott H & van Essen P 2005. 2003 Annual Report of the South Australian Birth Defects Register and incorporating the Annual report of Prenatal Diagnosis in South Australia, 2003, Women's and Children's Hospital Adelaide, South Australia.

Western Australia

Bower, C, Rudy E, Ryan A, Cosgrove P & Callaghan A ,2005. Report of the Birth Defects Registry of Western Australia 1980–2005, King Edward Memorial Hospital, Women's and Children's Health Service, Number 13.

Tasmania

Department of Health and Human Services 2005. Council of Obstetric and Paediatric Mortality and Morbidity, Tasmania: annual report for 2003. Hobart: Department of Health and Human Service.

Australian Capital Territory

Population Health Research Centre, ACT Health 2004. Maternal and perinatal health in the ACT 1997–2001. Canberra: ACT Government.

Appendix C: Data collection contacts for congenital anomalies

New South Wales

Ms Sue Travis
NSW Birth Defects Register
NSW Health Department
Locked Mail Bag 961
North Sydney NSW 2059
Phone: (02) 9391 9000
Fax: (02) 9391 9101
Email: nswhealth@doh.health.nsw.gov.au

Victoria

Ms Merilyn Riley
Victorian Births Defects Register
Victorian Perinatal Data Collection Unit
GPO Box 4003
Melbourne Vic 3001
Phone: (03) 9096 2695 or 1300 858 505
Fax: (03) 9096 2700
Email: perinatal.data@dhs.vic.gov.au

Queensland

Joanne Bunney
Data Services Unit
Health Information Centre
Queensland Department of Health
GPO Box 48
Brisbane Qld 4001
Phone: (07) 3234 1708
Fax: (07) 3234 0279
Email: joanne_bunney@health.qld.gov.au

South Australia

Ms Phillipa van Essen /Ms Heather Scott
The South Australian Birth Defects Register
Women's and Children's Hospital
72 King William Road
North Adelaide, SA 5006
Phone: (08) 8161 7368
Fax: (08) 8161 6088
Email: cywhs.sabdr@cywhs.sa.gov.au

Western Australia

Ms Edwina Rudy
The Western Australia Birth Defects Registry
King Edward Memorial Hospital
Women's and Children's Health Service
374 Bagot Road
Subiaco WA 6008
Phone : (08) 9340 2735
Fax: (08) 9340 2636
Email: Edwina.Rudy@health.wa.gov.au

Tasmania

MS Helen Galea

Clinical Data Services

Hospitals and Ambulance Service

Department of Health and Human Services

Level 2, 10 Murray Street

Hobart Tas 7000

Phone: (03) 6233 2173

Fax: (03) 6233 3550

Australian Capital Territory

Ms Maureen Bourne

Population Health Research Centre

ACT Health

Building 5, Level 1

The Canberra Hospital

POBox 11

Woden ACT 2606

Phone: (02) 6205 2612

Fax: (02) 6244 4138

Northern Territory

Acute Care Information Services

Department of Health and Community Services

Building 6, Royal Darwin Hospital

PO Box 41326

Casuarina NT 0810

Phone: (08) 8922 7779

Fax: (08) 8922 7787

Australian Paediatric Surveillance Unit

The Australian Paediatric Surveillance Unit

The Children's Hospital, Westmead

Level 2, Clinical Sciences Building

Locked Bag 4001,

Westmead NSW 2145

Phone: (02) 9845 3005/2200

Fax: (02) 9845 3082

Email: apsu@chw.edu.au

Appendix D: Committees

National Congenital Anomalies Steering Committee

New South Wales

Associate Professor Elizabeth Elliott
Director
Australian Paediatric Surveillance Unit
Department of Paediatrics and Child Health
University of Sydney

Dr Lee Taylor
Manager
Surveillance Methods
Centre for Epidemiology and Research
NSW Department of Health

Associate Professor Bridget Wilcken
Clinical Director
NSW Biochemical Genetics and
Newborn Screening Services

Ms Kerry Innes
Associate Director
Classification Support and
Development Division
Faculty of Health Sciences
University of Sydney

Dr. Hugh Martin
Paediatric Surgeon
Royal Australasian College of
Paediatric Surgeons
Children's Hospital, Westmead

Victoria

Associate Professor Jane Halliday
Epidemiologist
Birth Defects Register
Perinatal Data Collection Unit
Victorian Department of Human Services

Ms Christine Stone
Budget, Review and Finance Branch
Victorian Department of Human Services

Queensland

Professor David Tudehope
Director of Neonatology
Mater Mothers' Hospital

Ms Sue Cornes
Senior Director
Health Information Centre
Queensland Health

South Australia

Dr Anabelle Chan
Senior Medical Consultant
Pregnancy Outcome Unit
South Australian Health Commission

Western Australia

Professor Carol Bower
Principal Research Fellow
Head, Division of Population Sciences
The University of Western Australia

Tasmania

Mr Peter Mansfield
Manager
Clinical Data Services
Department of Health and Human Services

Australian Capital Territory

Ms Maurine Bourne
Data Manager
Population Health Research Centre
ACT Health

Northern Territory

Ms Sonya McNellee
Acute Care Information Services
Department of Health and Community Services

AIHW

The Unit Head
Children Youth and Families Unit
Australian Institute of Health and Welfare

Abbreviations and Symbols

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
AMC	Arthrogryposis multiplex congenita (congenital arthrogyposis)
APSU	Australian Paediatric Surveillance Unit
ASCCSS	Australian Standard Classification of Countries for Social Statistics
ASGC	Australian Statistical Geographical Classification
BPA	British Paediatric Association
CHARGE	coloboma, heart disease, atresia of choanae, retarded mental development and growth, genital hypoplasia, ear abnormalities-deafness
DSS	Data Set Specification
FAS	Fetal alcohol syndrome
FSANZ	Food Standards Australia New Zealand
g	grams
HDSC	Health Data Standards Committee
HGSA	Human Genetics Society of Australasia
ICBDSR	International Clearinghouse for Birth Defects Surveillance and Research
ICD	International Classification of Diseases
ICD-9	International Classification of Diseases, 9th Revision
BPA Classification of Diseases	British Paediatric Association Classification of Diseases
ICD-9-CM	International Classification of Diseases, 9th Revision, Clinical Modification
ICD-10-AM	International Statistical Classification of Diseases and Related Health Problems, 10th Revision, Australian Modification
METeOR	Metadata online registry
NCASC	National Congenital Anomalies Steering Committee
NCC	National Coding Centre
NCCH	National Centre for Classification in Health
NHDD	National Health Data Dictionary
NHIMPC	National Health Information Management Principal Committee
NHMRC	National Health and Medical Research Council
NMDS	National Minimum Data Set

NPDC	National Perinatal Data Collection
NPDDC	National Perinatal Data Development Committee
NPSU	National Perinatal Statistics Unit
NSW	New South Wales
NT	Northern Territory
NTD	Neural tube defect
PWS	Prader-Willi syndrome
Qld	Queensland
RACP	Royal Australian College of Physicians
SA	South Australia
SACC	Standard Australian Classification of Countries
SIMC	Statistical Information Management Committee
SLA	Statistical Local Area
STICCA	State and Territory Implementation Committee for Congenital Anomalies
Tas	Tasmania
TOP	Termination of pregnancy
UNSW	University of New South Wales
Vic	Victoria
WA	Western Australia
WHO	World Health Organization
n.p.	Not published
. .	Not applicable

Glossary

Birthweight: the first weight of the baby (stillborn or live born) obtained after birth (usually measured to the nearest 5g and obtained within one hour of birth).

Congenital anomalies: a structural, functional or metabolic abnormality that is present at birth, even if not diagnosed until months or years later.

Fetal death (stillbirth): death prior to the complete expulsion or extraction from its mother of a product of conception of 20 or more completed weeks of gestation or of 400g or more birthweight. The death is indicated by the fact that after such separation the fetus does not breathe or show any other evidence of life, such as beating of the heart, pulsation of the umbilical cord, or definite movement of voluntary muscles.

Gestational age: the duration of pregnancy in completed weeks calculated from the date of the first day of a woman's last menstrual period and her baby's date of birth, or via ultrasound, or derived from clinical assessment during pregnancy or from examination of the baby after birth.

Termination of pregnancy: termination of pregnancy (induced abortion) by medical or mechanical means before 20 weeks of gestation.

Infant death: death of a live birth child under 1 year of age. Infant death rates are expressed per 10,000 live births in this report.

Isolated malformation: only one major congenital anomaly is present in the infant or fetus.

Live birth: the complete expulsion or extraction from its mother of a product of conception, irrespective of the duration of the pregnancy, which, after such separation, breathes or shows any other evidence of life, such as beating of the heart, pulsation of the umbilical cord, or definite movement of voluntary muscles, whether or not the umbilical cord has been cut or the placenta is attached; each product of such a birth is considered live birth.

Low birthweight: birthweight of less than 2,500 g.

Major congenital anomaly: congenital anomaly that are either lethal or significantly affect the individual's function or appearance.

Maternal age: mother's age at the birth of her baby.

Multigravida: pregnant woman who has had at least one previous pregnancy.

Multipara: pregnant woman who has had at least one previous pregnancy resulting in a live birth or stillbirth.

Neonatal death: death of a live born baby within 28 days of birth.

Parity: number of previous pregnancies resulting in live births or stillbirths, excluding the current pregnancy.

Perinatal death: a fetal or neonatal death of at least 20 weeks gestation or at least 400g birthweight.

Plurality: the number of births resulting from a pregnancy.

Post-term birth: birth at 42 or more completed weeks of gestation.

Preterm birth: birth before 37 completed weeks of gestation.

Primipara: pregnant woman who has had no previous pregnancy resulting in a live birth or stillbirth.

Stillbirth: see Fetal death.

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