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Congenital Malformations Australia 1993 and 1994

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Requests for data

Any enquiries about data for regions within States and Territories and for individual hospitals should be directed initially to State and Territory health departments. Other enquiries should be made to the address below.

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Abbreviations

NSW	-	New South Wales
Vic	-	Victoria
Qld	-	Queensland
WA	-	Western Australia
SA	-	South Australia
Tas	-	Tasmania
ACT	-	Australian Capital Territory
NT	-	Northern Territory
ABS	-	Australian Bureau of Statistics
AIHW	-	Australian Institute of Health and Welfare
NPSU	-	National Perinatal Statistics Unit
a/s	-	atresia/stenosis
ASD	-	atrial septal defect
BPA	-	British Paediatric Association
bronch.	-	bronchus
cerv.	-	cervix
dis.	-	disease
dysgen.	-	dysgenesis
grt.	-	great
ICD	-	International Classification of Diseases
L	-	left
musculoskelet.	-	musculoskeletal
NEC	-	not elsewhere classified
NOS	-	not otherwise specified
Pat.	-	patent
pulm.	-	pulmonary
spec.	-	specified
st./insuffic.	-	stenosis/insufficiency
synd.	-	syndrome
TOF	-	tracheo-oesophageal fistula
trach.	-	trachea
transpstn.	-	transposition
Vent.	-	ventricular

Highlights

- Among 3.37 million births in the period from 1981 to 1994, 53,065 (1.6%) infants with major congenital malformations diagnosed at birth or in the first 28 days were notified to the national monitoring system. These included 4,426 (1.7%) infants born in 1993 and 4,297 (1.6%) in 1994.

- Perinatal deaths due to congenital malformations declined from 35.9 per 10,000 births in 1973 to 17.5 per 10,000 births in 1994, the lowest level of any year. The perinatal death rate for anencephalus showed the most pronounced decline but there were also substantial falls for spina bifida, hydrocephalus and congenital heart defects. Perinatal death rates due to chromosomal abnormalities increased in the same period. Congenital malformations accounted for 22% of perinatal deaths in 1994.

- Infant deaths due to congenital malformations declined from 28.8 per 10,000 live births in 1980 to 17.6 per 10,000 live births in 1994. Congenital malformations were the cause of 30% of infant deaths, and 10% of childhood deaths, in 1994.

- In the period from 1992 to 1994, the total malformation rate of 305.9 per 10,000 among births to mothers aged 40 years and over was almost double that of 154.8 per 10,000 among births to mothers aged 20-24 years. Chromosomal abnormalities were more likely with advancing maternal age. Women aged 40 years and over were 12 times more likely than women aged 20-24 years to give birth to an infant with a chromosomal abnormality and more than 60 times more likely to have a termination of pregnancy before 20 weeks' gestation.

- Isolated and multiple malformations were more common in twins and other multiple births than in singleton births.

- Following a marked decline in the rate of anencephalus among births up to 1992, there was a slight increase in 1993 but then a continuation of the downward trend in 1994. Spina bifida also declined to its lowest rate of 2.9 per 10,000 births in 1994. In that year, there were 84 reported terminations of pregnancy for anencephalus and 70 for spina bifida, the largest numbers in any year.

- Increasing rates of ventricular septal defect and obstructive defects of the renal pelvis and ureter are likely to reflect better ascertainment, particularly by prenatal ultrasound screening for the renal defects.

- The rate of gastroschisis reached its highest level of 2.0 per 10,000 births in 1993 and then declined to 1.5 per 10,000 births in 1994. This malformation is much commoner among births to younger than to older mothers.

- Notified terminations of pregnancy for fetal malformations increased from 421 in 1991 to 718 in 1994, but notification was incomplete. The main indications for termination of pregnancy were trisomy 21 (Down syndrome), other chromosomal abnormalities and neural tube defects (anencephalus and spina bifida). Most terminations of pregnancy for fetal malformations (75.7%) were performed at gestational ages between 16 and 22 weeks.

- The number of reported terminations of pregnancy for trisomy 21 (Down syndrome) and for trisomy 18 (Edwards syndrome) was higher in 1994 than in any previous year - 128 and 48, respectively.

1 Introduction

Congenital malformations are a significant public health problem because they are relatively common, they are major reasons for admission to hospital during infancy and childhood, they may frequently lead to disabilities and handicaps, and some types are fatal. In 1994, 21.8% (452/2,076) of all perinatal deaths in Australia, and 30.0% (454/1,512) of infant deaths, were due to congenital malformations (Australian Bureau of Statistics 1995).

Data collected in the State and Territory perinatal data systems, by birth defects registers, and from hospitals provide valuable information on the number of children with malformations diagnosed early in life. These data enable analysis of trends over time and variations by region; collaborative studies of the descriptive epidemiology of congenital malformations, both within Australia and internationally; and evaluation of the impact of prenatal diagnosis or other interventions on trends in occurrence. Almost universal prenatal screening of pregnant women by ultrasound in Australia, and the frequent use of amniocentesis or chorionic villus sampling in high-risk women, has increased the likelihood of detecting some types of congenital malformations during fetal life. If a pregnancy is terminated, it is important to collect information about these pregnancies so that the effect on trends can be monitored adequately.

This report contains national data on congenital malformations among births that occurred in the period 1981 to 1994. Data for the years up to 1992 were published previously in *Birth Defects Series Number 1* (Lancaster & Pedisich 1995).

1.1 Criteria

Congenital malformations are anatomical defects or chromosomal abnormalities that are present at birth. Major congenital malformations are either lethal or significantly affect the individual's function or appearance. Minor malformations do not have functional or cosmetic importance but may sometimes signify an underlying genetic disorder. Although infants with only minor malformations may be included in State birth defects registers, these infants are not notified to the AIHW National Perinatal Statistics Unit unless major malformations are also present (see Appendix 1 for list of minor malformations).

Major congenital malformations diagnosed in liveborn infants in the first 28 days, or in stillbirths of at least 20 weeks' gestation or 400g birthweight, are included in this report. Some tables on selected malformations also give incomplete data on terminations of pregnancy that followed prenatal diagnosis. If terminations of pregnancy occurred at gestational ages of 20 weeks or more, these fetuses are included in the figures for stillbirths.

1.2 Sources of data

Congenital malformations are notified on forms designed to obtain data on all births occurring in each State and Territory. Ascertainment of congenital malformations is improved by using additional sources of notification including children's hospitals and other referral hospitals, cytogenetics laboratories, perinatal death certificates, autopsy reports and notifications of induced abortions. Four States - New South Wales, Victoria, Western Australia and South Australia - have birth defects registers that include notifications of malformations diagnosed in infants and children after the perinatal period.

Only South Australia has mandatory notification of induced abortions. Ascertainment of terminations of pregnancy that follow prenatal diagnosis of fetal abnormalities is variable in the other States and Territories, except for chromosomal abnormalities which have been reported by cytogenetics laboratories since 1982.

Data for 1981 were incomplete in New South Wales and excluded births in Victoria, Western Australia, the Australian Capital Territory and the Northern Territory. Notifications of congenital malformations in the Northern Territory began in 1986.

1.3 Data items

Demographic, maternal and infant or fetal data are recorded on each notification to the AIHW National Perinatal Statistics Unit (NPSU). The data items are listed in Appendix 2. Some items are incomplete, either because they are not recorded on notification forms or because the information may not be readily available at the data source (eg children's hospitals, cytogenetics laboratories).

South Australia is the only State that routinely collects some information on the family history of birth defects or on maternal exposures to drugs and environmental agents. Such information is not coded in the national monitoring system but has been used to review reported associations between specific drugs (eg isotretinoin) and congenital malformations.

1.4 Data processing

Data on congenital malformations are coded in each State and Territory by the groups responsible for perinatal data collection and birth defects registers. Major congenital malformations and chromosomal abnormalities listed in the chapter on congenital anomalies in the Ninth Revision of the International Classification of Diseases (ICD) are notified to the NPSU. Both the NPSU and some States use the British Paediatric Association (BPA) Classification of Diseases, which is a 5-digit system compatible with ICD at the 4-digit level, to code congenital malformations. The BPA classification enables more specific coding of malformations and, with some modification of codes for malformation syndromes, limb reduction defects and other selected malformations, has been used by the NPSU since 1981. The list of minor malformations that are excluded from the national data are given in Appendix 2. If notification forms contain information on which side of the body is affected by a malformation, this information is coded by the NPSU.

The State and Territory groups send copies of notifications in regular batches to the NPSU throughout the year. Since 1994, some States have provided data in an agreed format on floppy disk. Whenever possible, coding by State and Territory groups and the NPSU is based on written descriptions of congenital malformations. Further information is requested if diagnoses are doubtful or if key data items such as maternal age are missing. While this report gives widely accepted definitions of selected major congenital malformations, it is not always possible to ensure that notified diagnoses of malformations conform to these definitions.

1.5 Contents of report

Data in all tables, graphs and maps are based on the year of birth, or the year of induced abortion, and on the State or Territory in which the birth or termination of pregnancy occurred. Denominators for calculating rates are based on the State or Territory of registration, obtained from the Australian Bureau of Statistics. Malformation rates are expressed per 10,000 total births, including those malformations that are specific to one sex. As the criteria used by the States and Territories for their published reports may differ from those used here, their numbers and rates may differ from those in this report. Also, the criteria and sources of notifications vary among the States and Territories, so differences in malformation rates should be interpreted cautiously. Small numbers of specific types of malformations may also influence variations in rates.

Tables 2.1-2.7 are similar in content to annual tables published previously. Tables 2.1-2.4 give national data, and data for each State and Territory, on the total number of infants and fetuses with major congenital malformations in the years 1981 to 1994. In Tables 2.5-2.7, State/Territory and national data are given on major malformations affecting all anatomical systems. National data are presented separately for 1981-1992, 1993, 1994 and the whole period; data on selected malformations in each State and Territory are given for the same years.

Tables 3.1-3.50 and the accompanying graphs and maps (Figure 3.1-3.50), provide data on national trends and variations by State and Territory, of 25 congenital malformations or chromosomal abnormalities that are either lethal, have significant consequences for surviving children and their

families, or are relatively common. Further information of the descriptive epidemiology of these malformations was given in the previous report (Lancaster & Pedisich 1995).

As the level of ascertainment of induced abortions at less than 20 weeks' gestation is uncertain in most States and Territories, these abortions have been excluded from the reported rates of the 25 selected malformations. Induced abortions of fetuses less than 20 weeks' gestation, or unstated gestational age, are given under that heading in the tables. Induced abortions at gestational ages of 20 weeks and over are included in the figures for stillbirths, but these terminations of pregnancy had not necessarily been reported as perinatal deaths. The inclusion of stillbirths in these Australian data will affect comparisons with those other countries where data on stillbirths are not available.

The data reported here on the proportion of stillborn and liveborn infants with selected malformations dying before birth or in the neonatal period (within 28 days of birth) may not always be reliable for several reasons. If a malformed infant dies of some related complication such as an infection or cardiac failure, the congenital malformation may not always be recorded on the perinatal death certificate. On the other hand, if there is incomplete reporting of malformations on birth notifications, the proportion of stillbirths and neonatal deaths may be overestimated because ascertainment of malformations recorded on perinatal death certificates is complete in all States and Territories. Also, because infants with multiple malformations are included in the tables for each type of malformation, there may be relatively high proportions of stillbirths and neonatal deaths, and sometimes induced abortions, included in the figures for some apparently mild malformations.

For each congenital malformation, the proportion occurring as an isolated malformation, in association with one or more other major malformations, or as part of a chromosomal syndrome is given. As ascertainment of major malformations has generally continued to improve in the States and Territories in recent years, comparative data by State and Territory of birth are presented for the five-year period of 1990 to 1994. For some of the less common malformations, these comparisons are affected by the relatively small number of births in some States and Territories. It is apparent from examining these malformation rates that even more striking variations are sometimes likely when areas with fewer births in shorter time periods are considered.

During the period from 1982 to 1994, the annual number of births increased from 238,684 in 1982 (excluding the Northern Territory) to 261,335 in 1994 with some fluctuations in the intervening years (Table 2.19).

1.6 International monitoring of congenital malformations

National, regional or hospital-based monitoring systems similar to the Australian national system operate in numerous other countries around the world. Through the International Clearinghouse for Birth Defects Monitoring Systems, and its International Centre for Birth Defects located in Rome, Australia participates in quarterly and annual reporting of congenital malformations and in studies of the epidemiology and causes of congenital malformations. The definitions of selected major congenital malformations in this report are generally those adopted by the International Clearinghouse for Birth Defects Monitoring Systems.

2 Major congenital malformations

2.1 Births

There were 53,065 infants and fetuses with major congenital malformations notified in the years 1981 to 1994 among 3.37 million births, a total rate of 157.6 per 10,000 births, or 1.6%, in this 14-year period (Table 2.1). The total malformation rate of 164.4 per 10,000 births in 1994 was slightly less than the rate of 169.9 per 10,000 in 1993. Overall, 76.5% of infants had malformations affecting a single body system, 7.4% had multiple malformations affecting more than one system, and 16.2% had identifiable chromosomal or other syndromes.

The reported malformation rates in the 5-year period from 1990 to 1994 were highest in Victoria (215.1 per 10,000 births), Queensland (189.1 per 10,000 births) and South Australia (184.0 per 10,000 births) and lowest in the Australian Capital Territory (99.5 per 10,000 births) and Tasmania (112.9 per 10,000 births) (Table 2.2). These variations are likely to reflect differences in the sources and ascertainment of malformations rather than real differences in incidence.

Each State and Territory publishes data on congenital malformations, either in separate reports on birth defects (Bower et al. 1996; Riley & Halliday 1996; Taylor et al. 1996; The South Australian Birth Defects Register 1996) or in reports on all births (Ascroft 1992; Markey et al. 1996; Marsden; Queensland Health 1996). The malformation rates in this report may differ from rates published by the States and Territories because of differences in the age criteria for inclusion of infants, differences in the criteria for including major and minor malformations and other birth defects, varying sources of data, and occasionally differences in coding practices. Comparisons of some congenital heart defects and other malformations diagnosed beyond the perinatal period are particularly affected by these factors.

The major source of notifications was the perinatal data collected on all births in each State and Territory (Table 2.3). Other important sources were perinatal death certificates and sometimes autopsy reports, and reports of chromosomal abnormalities from cytogenetics laboratories. Notifications from children's hospitals in New South Wales were no longer specified in the data after 1992, accounting for the sharp decline in the proportion of notifications from that source.

The main anatomical systems in which major malformations occurred were the musculoskeletal and cardiovascular systems and genital organs (Table 2.4). The specific malformations contributing to these different systems are shown for Australia for births in 1981 to 1992, 1993 and 1994 (Table 2.5) and for each State and Territory (Tables 2.6, 2.7).

Comparison of total malformation rates by year and by State and Territory may be influenced both by the completeness of clinical detection and notification of major malformations and by the extent to which the various sources of notifications are used. The ascertainment of three relatively common malformations - congenital dislocation of the hip, ventricular septal defect, and hypospadias - may vary considerably, affecting total malformation rates. Congenital dislocation of the hip accounted for more than 60% of musculoskeletal malformations, ventricular septal defect was the most frequently notified congenital heart defect, and 81% of malformations of the genital organs were due to hypospadias.

Most of the difference between the total malformation rates of 169.9 per 10,000 births in 1993 and 164.4 per 10,000 in 1994 could be attributed to these three malformations. The reported rate of ventricular septal defect decreased by 0.7 per 10,000, hypospadias by 0.6 per 10,000, and congenital dislocation of the hip by 3.0 per 10,000 between 1993 and 1994 (Table 2.5). Similarly, the reported rate of ventricular septal defect in 1990-1994 varied threefold from 6.7 per 10,000 in the Australian Capital Territory and 7.5 per 10,000 in Tasmania to 23.1 per 10,000 in the Northern Territory and 23.2 per 10,000 in Victoria (Table 2.7). There were even greater variations in the reported rates of hypospadias in the same period, from 3.4 per 10,000 in the Northern Territory to 27.6 per 10,000 in Victoria. The reported rate of congenital dislocation of the hip varied between 0.4 per 10,000 births in the Australian

Capital Territory and 43.2 per 10,000 in Queensland. These findings emphasise that the variations in total malformation rates, and in the rates of specific malformations known to have varying ascertainment, should be interpreted with caution.

The source of notification of malformations affects the completeness of reporting of various maternal and infant variables. When information is obtained from death certificates, cytogenetics laboratories or children's hospitals, missing data are more likely than when the perinatal data form is the source. The proportion with missing information among births notified to the NPSU varied between 0.4% for the infant's sex and 25.8% for the maternal country of birth (Table 2.8). The completeness of reporting of these variables can be improved by linking notifications from other sources with the perinatal data form for each birth.

2.2 Terminations of pregnancy

South Australia is the only State that has a legislative requirement for notification of terminations of pregnancy performed for any indication, including terminations after prenatal diagnosis of congenital malformations (Chan & Taylor 1991). Birth defects registers in New South Wales, Victoria, South Australia and Western Australia obtain information on termination of pregnancy and ascertainment has improved considerably in the past decade. Also, although the other States and Territories do not have birth defects registers, information on some terminations has been provided from cytogenetics laboratories and sometimes from other sources. All States and Territories provide the available information on terminations of pregnancy to the AIHW National Perinatal Statistics Unit, but the level of completeness of national data remains uncertain. By comparing the national trends, and the numbers reported by each State and Territory, some inferences can be drawn about the overall level of reporting.

For this report, terminations of pregnancy for fetal malformations are divided into two main groups - induced abortions performed at less than 20 weeks' gestation (or when gestational age was not stated) and induced births at 20-27 weeks' gestation. The latter group includes fetuses that have reached a gestational age of 20 weeks, at which registration of perinatal deaths and notification in State and Territory perinatal collections is required. If there is information indicating that these terminations occurred after prenatal diagnosis, this is recorded for each notification so that a distinction can be made between induced births and other non-induced stillbirths occurring at the same gestational age of 20-27 weeks.

In the four-year period from 1991 to 1994, the reported number of induced abortions increased from 325 in 1991 to 579 in 1994 (Table 2.9). There were relatively fewer induced births, increasing from 96 in 1991 to 139 in 1994. The induced births accounted for 22.8% of all reported terminations of pregnancy performed for fetal malformations in 1991 and 19.4% in 1994.

The most common indication for terminations of pregnancy were trisomy 21, other chromosomal abnormalities, and neural tube defects (anencephalus, spina bifida and encephalocele) (Table 2.9). Fetuses with multiple malformations are enumerated for each specific type of malformation and the numbers of malformed fetuses with single or multiple malformations, or malformation syndromes, are also given.

To show trends in the notified terminations and their relationship to births in the perinatal collections, both induced abortions and induced births are expressed as ratios per 10,000 births. The ratio of notified induced abortions for all fetal malformations increased from 12.7 per 10,000 births in 1991 to 22.2 per 10,000 in 1994. Reflecting their relatively smaller numbers, the ratio of notified induced births rose from 3.7 per 10,000 births in 1991 to 5.3 per 10,000 in 1994. The overall ratio of notified terminations of pregnancy for fetal malformations increased from 16.4 per 10,000 births in 1991 to 27.5 per 10,000 in 1994, indicating that there was about 1 termination for every 360 births in 1994. This rising trend in terminations is attributable to improving ascertainment and also to a real increase in terminations performed for chromosomal abnormalities, which have been well reported for more than a decade, and possibly other malformations.

The overall increase in terminated pregnancies between 1991 and 1994 does not appear to be due to any marked change in the relative number of induced abortions performed at gestational ages of less than 16 weeks or more than 22 weeks (Table 2.10, Figure 2.1). The duration of pregnancy was not given for 18.9% of notified terminations in this period. Among terminations with known gestational ages, three-quarters (75.7%) were performed between 16 and 22 weeks and the modal week was either 18 or 19 weeks in different years.

Chromosomal abnormalities, particularly trisomy 21, accounted for the majority of terminations performed before 16 weeks (Table 2.11, Figure 2.2.). Terminations for anencephalus, spina bifida and other malformations detected by ultrasound or other methods of prenatal diagnosis were more likely to occur just before 20 weeks, when most pregnant women are screened by ultrasound examination.

2.2.1 Notifications of terminations of pregnancy in each State and Territory

As already noted, there is varying ascertainment of induced abortions and induced births in different States and Territories. Also, additional information from autopsy reports provided with notifications of terminations to the NPSU enables coding of prenatal diagnostic methods and the indications for terminations, particularly for those at gestational ages of 20 weeks or more. Active review of hospital records by staff of birth defects registers in some States, particularly in Victoria, is probably an important factor influencing comparisons of terminations between the States where there is no legal requirement to notify terminations.

The reported numbers of terminations are affected by these various factors and should not be regarded as the complete figures for any State or Territory (Table 2.12). Nevertheless, by comparing the reported numbers from the different States and Territories, valuable insights can be obtained for improving ascertainment and for reducing the deficiencies of the national data.

By comparing the ratios of terminations for the most recent year's data (1994), the effect of improving ascertainment on differences in ratios can be avoided to some extent. In 1994, for terminations of pregnancy at less than 20 weeks (including unstated gestational ages), the highest reported ratios were in Western Australia (35.8 per 10,000 births) and Victoria (34.7 per 10,000 births) and the lowest was in Queensland (3.3 per 10,000 births) (Table 2.13). For terminations at 20-27 weeks, the reported ratios in South Australia (18.2 per 10,000 births) and the Australian Capital Territory (16.7 per 10,000 births) were higher than in the other States. When data on induced abortions and induced births are combined, the highest reported ratios of terminations were in South Australia (47.5 per 10,000 births), the Australian Capital Territory (46.0 per 10,000 births) and Victoria (44.4 per 10,000 births), and the lowest ratio was in Queensland (4.2 per 10,000 births).

The gestational age distribution of the terminations of pregnancy showed considerable variation in the different States (Table 2.14, Figure 2.3). Because of small numbers, the data for Tasmania, the Australian Capital Territory and the Northern Territory were combined as 'other'. Relatively more terminations reported from Victoria and South Australia were induced births at 20-27 weeks.

As the forms used to obtain information about terminations of pregnancy often have limited data on maternal characteristics, there are substantial deficiencies in some variables such as maternal country of birth and Indigenous status. These variables may be important in analysing differences in malformation rates between the various population groups. In all States and Territories, a high proportion of notified induced abortions at less than 20 weeks lacked this information (Table 2.15). Also, the gestational age was not reported for 24.9% of these terminations. This information is needed to analyse the impact of different methods of prenatal diagnosis. For example, chorionic villus sampling is usually performed at an earlier stage of pregnancy than amniocentesis in screening for chromosomal abnormalities. Variations in the relative use of these two tests and the gestational ages at which they are done will affect interpretation of differences in rates.

2.3 Congenital malformations by maternal age

In the three-year period from 1992 to 1994, malformation rates among births were generally higher for younger and older mothers (Table 2.16). There was more variation by maternal age among infants with multiple malformations than for those with an isolated malformation. There was a pronounced association between advancing maternal age and an increasing rate of chromosomal abnormalities, ranging from 9.5 per 10,000 births for infants of mothers aged less than 20 years to 146.0 per 10,000 births for infants whose mothers were 40 years and over. Except for higher rates in the youngest and oldest maternal age groups, other non-chromosomal syndromes did not vary much with maternal age. The previous report on *Congenital Malformations Australia 1981-1992* (Lancaster & Pedisich 1995) gave data on the maternal age distribution of 25 selected major malformations.

For terminations at gestational ages of less than 20 weeks, the ratio of syndromes due to chromosomal abnormalities increased with advancing maternal age. Isolated malformations were also more likely with advancing maternal age but this association was much less marked than for chromosomal abnormalities. There was no clear association between maternal age and multiple malformations or non-chromosomal syndromes.

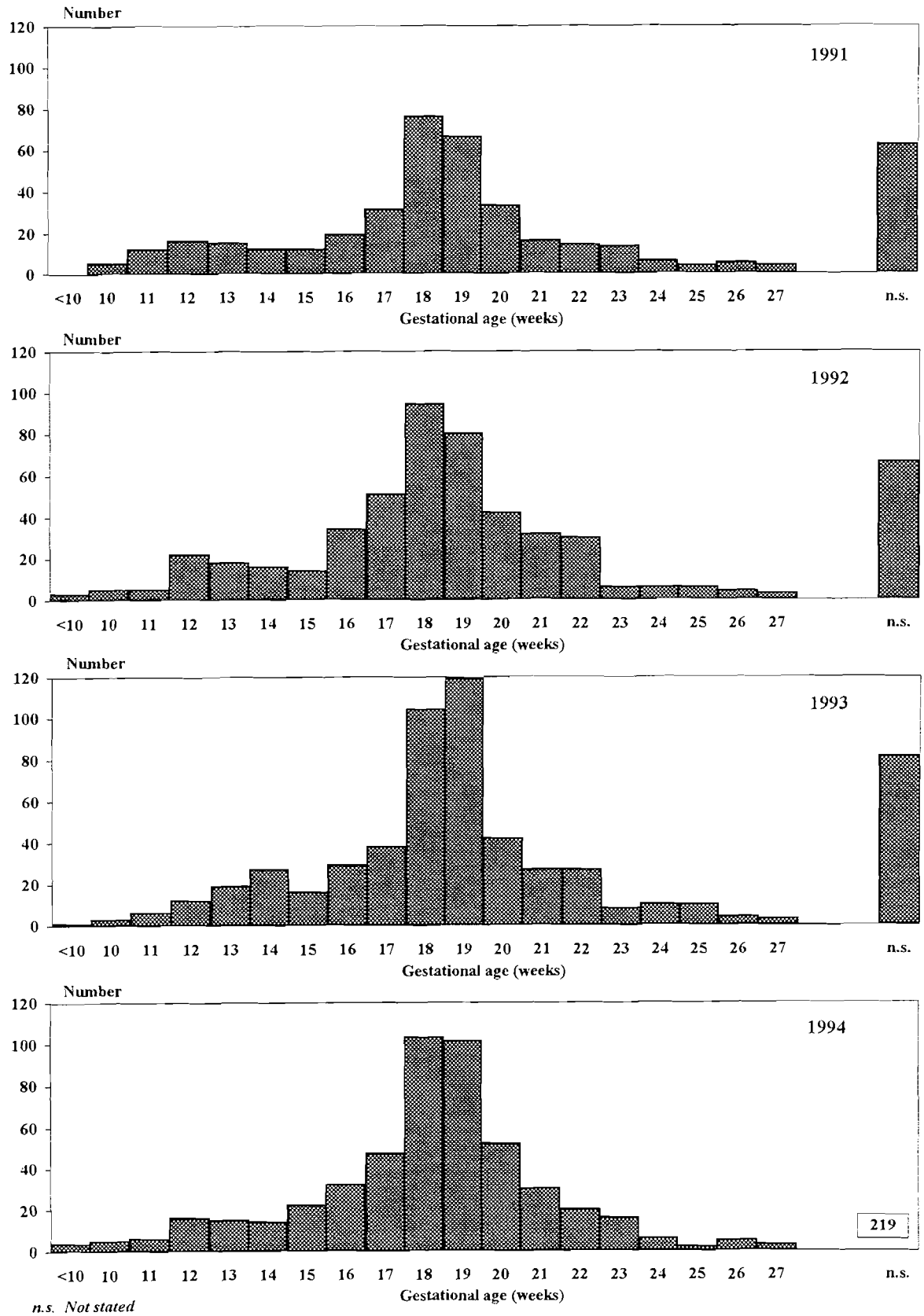
2.4 Congenital malformations in singleton and multiple births

In 1992 to 1994, there were higher rates of isolated and multiple malformations in twins and other multiple births than in singleton births (Table 2.17). Singleton infants and twins had similar rates of chromosomal abnormalities, but there were higher rates of non-chromosomal syndromes in twins than in singleton births. Relatively few terminations for fetal malformations were performed in multiple pregnancies, so little is gained by comparing them with singleton pregnancies.

2.5 Selected congenital malformations by maternal country of birth

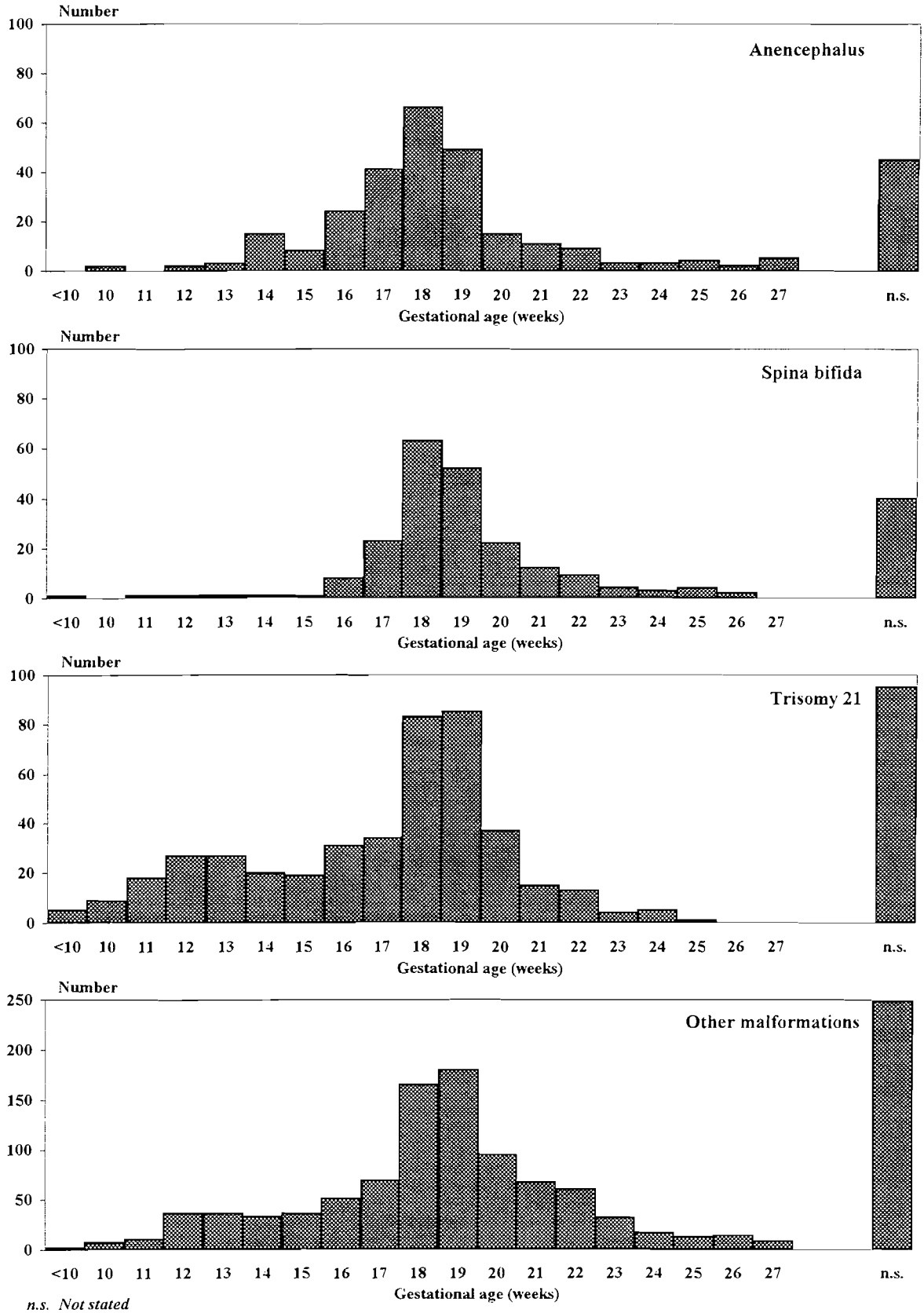
Maternal country of birth is usually recorded in the perinatal data collections but this information is often lacking for other sources of notification of major congenital malformations, especially for terminations of pregnancy before 20 weeks (Table 2.15), perinatal deaths and notifications from cytogenetic laboratories. For births, this deficiency can be overcome by linking the various sources of notification with the perinatal data, but such linkage is not available for terminations before 20 weeks. Any comparison of malformation rates between countries could also be affected by differences in maternal age distribution, by differences in access to prenatal diagnostic services, by varying cultural attitudes to termination of pregnancy, as well as by the relatively small number of births for some countries. These factors need to be considered in interpreting variations in malformation rates among births to mothers born in different countries and to Indigenous and non-Indigenous mothers (Table 2.18).

Figure 2.1: Terminations of pregnancy for fetal malformations, by gestational age, Australia, 1991-1994



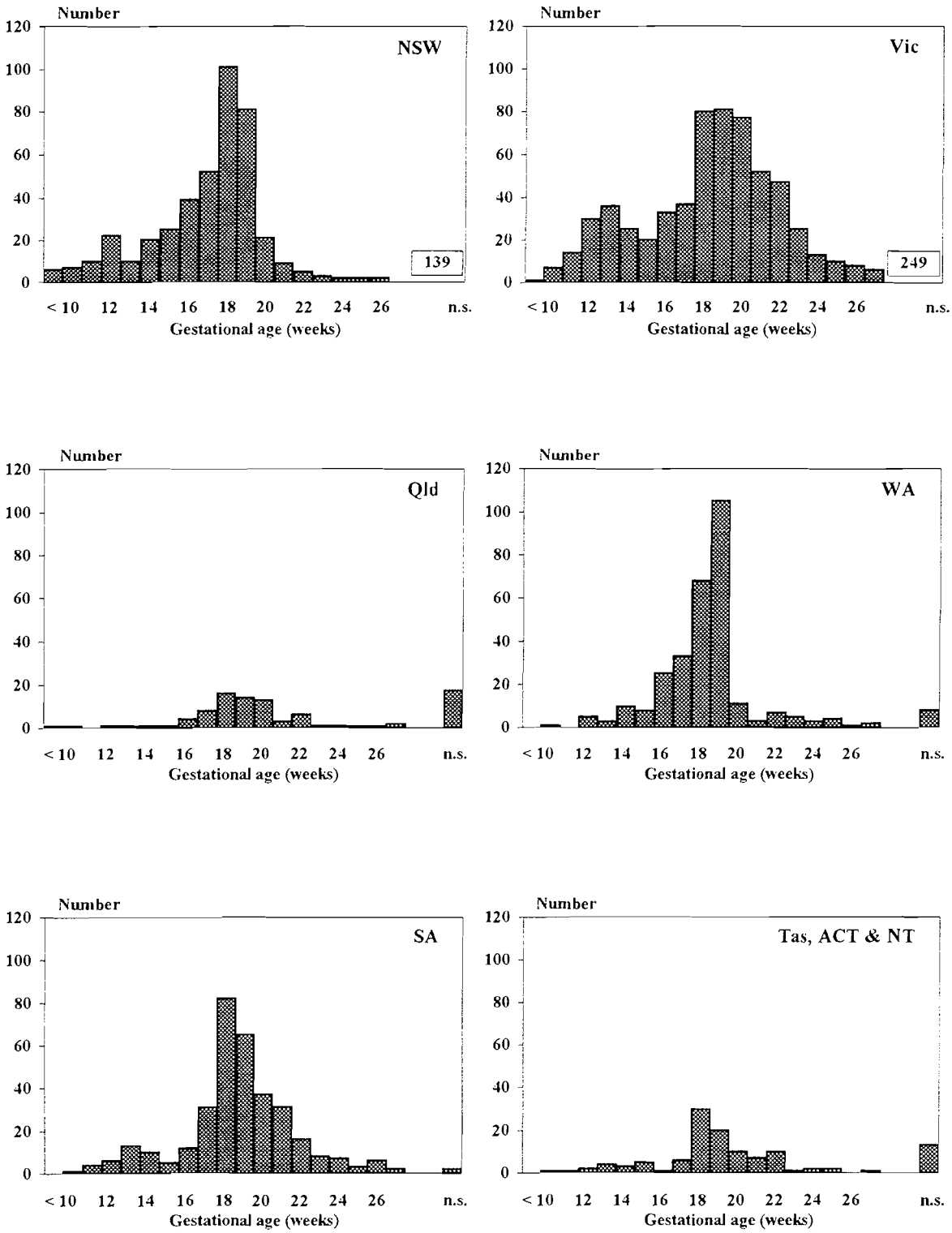
n.s. Not stated

Figure 2.2: Terminations of pregnancy for fetal malformations, by type of malformation, Australia, 1991-1994



n.s. Not stated

Figure 2.3: Terminations of pregnancy for fetal malformations, by gestational age, selected States, 1991-1994



n.s. Not stated

Table 2.1: *Single and multiple congenital malformations, Australia, 1981-1994*

Type of malformation	1981-92	1993	1994	1981-94	1981-92	1993	1994	1981-94
	Number				Rate per 10,000 births			
All types	44,342	4,426	4,297	53,065	155.9	169.9	164.4	157.6
Single system	33,967	3,365	3,253	40,585	119.4	129.1	124.5	120.5
Multiple systems	3,323	261	318	3,902	11.7	10.0	12.2	11.6
- 2 systems	2,236	179	232	2,647	7.9	6.9	8.9	7.9
- 3+ systems	1,038	82	81	1,201	3.6	3.1	3.1	3.6
- unknown	49	-	5	54	0.2	-	0.2	0.2
Syndrome	7,052	800	726	8,578	24.8	30.7	27.8	25.5

Note: Data for 1981 exclude Vic, WA, ACT, NT and certain hospital in NSW; data for 1982-1985 exclude NT.

Table 2.2: *Single and multiple congenital malformations by State or Territory of birth, 1990-1994*

Type of malformation	NSW	Vic	Qld	WA	SA	Tas	ACT	NT	Australia
					Number				
All types	6,861	7,072	4,380	1,564	1,836	392	236	223	22,564
Single system	5,026	5,649	3,580	1,101	1,431	282	160	161	17,390
Multiple systems	538	395	183	140	139	20	9	24	1,448
- 2 systems	376	296	136	91	100	14	7	15	1,035
- 3+ systems	152	96	45	47	39	5	2	9	395
- unknown	10	3	2	2	-	1	-	-	18
Syndrome	1,297	1,028	617	323	266	90	67	38	3,726
					Rate per 10,000 births				
All types	155.0	215.1	189.1	123.6	184.0	112.9	99.5	125.5	172.8
Single system	113.6	171.8	154.5	87.0	143.4	81.2	67.5	90.6	133.2
Multiple systems	12.2	12.0	7.9	11.1	13.9	5.8	3.8	13.5	11.1
- 2 systems	8.5	9.0	5.9	7.2	10.0	4.0	3.0	8.4	7.9
- 3+ systems	3.4	2.9	1.9	3.7	3.9	1.4	0.8	5.1	3.0
- unknown	0.2	0.1	0.1	0.2	-	0.3	-	-	0.1
Syndrome	29.3	31.3	26.6	25.5	26.7	25.9	28.3	21.4	28.5

Table 2.3: *Source of notification of congenital malformations, Australia, 1990-1994*

Source	1990-92	1993	1994	1990-94	1990-92	1993	1994	1990-94
	Number				Per cent			
All sources	13,841	4,426	4,297	22,564	100.0	100.0	100.0	100.0
Birth notification	12,665	4,336	4,194	21,195	91.5	98.0	97.6	93.9
Referral hospital	1,331	-	31	1,362	9.6	-	0.7	6.0
Death certificate	1,079	220	193	1,492	7.8	5.0	4.5	6.6
Pathology report	604	163	160	927	4.4	3.7	3.7	4.1
Cytogenetics report	1,246	284	413	1,943	9.0	6.4	9.6	8.6

Table 2.4: *Congenital malformations by major anatomical system, Australia, 1981-1994*

Codes	Malformations	1981-92	1993	1994	1981-94	1981-92	1993	1994	1981-94
		Number				Rate per 10,000 births			
	All malformations	44,342	4,426	4,297	53,065	155.9	169.9	164.4	157.6
740-2	Nervous system	5,345	378	337	6,060	18.8	14.5	12.9	18.0
743	Eye	855	78	76	1,009	3.0	3.0	2.9	3.0
744	Ear, face & neck	313	44	48	405	1.1	1.7	1.8	1.2
745-6	Heart	8,123	888	914	9,925	28.6	34.1	35.0	29.5
747	Circulatory system	3,121	318	286	3,725	11.0	12.2	10.9	11.1
748	Respiratory system	824	78	58	960	2.9	3.0	2.2	2.9
749	Cleft palate/lip	4,196	402	382	4,980	14.7	15.4	14.6	14.8
750-1	Digestive system	3,281	303	270	3,854	11.5	11.6	10.3	11.4
752	Genital organs	6,892	664	626	8,182	24.2	25.5	24.0	24.3
753	Urinary system	3,667	433	514	4,614	12.9	16.6	19.7	13.7
755	Limbs	3,938	418	379	4,735	13.8	16.0	14.5	14.1
754, 6	Other musculoskeletal	10,075	902	801	11,778	35.4	34.6	30.7	35.0
757	Integument	311	35	66	412	1.1	1.3	2.5	1.2
758	Chromosomal	5,449	636	604	6,689	19.2	24.4	23.1	19.9
759	Other & unspecified	1,125	129	104	1,358	4.0	5.0	4.0	4.0
760	Maternal conditions	53	2	2	57	0.2	0.1	0.1	0.2

Note: Data for 1981 exclude Vic, WA, ACT, NT and certain hospital in NSW; data for 1982-1985 exclude NT.

Table 2.5: *Selected congenital malformations, Australia, 1981-1994*

Codes	Malformations	1981-1992	1993	1994	1981-94	1981-1992	1993	1994	1981-94
		Number				Rate per 10,000 births			
740	Anencephalus & similar anomalies	1,127	64	44	1,235	4.0	2.5	1.7	3.7
740.0	Anencephalus	962	57	42	1,061	3.4	2.2	1.6	3.2
740.1	Craniorachischisis	148	6	1	155	0.5	0.2	0.0	0.5
740.2	Iniencephaly	18	1	1	20	0.1	0.0	0.0	0.1
741	Spina bifida	1,834	103	77	2,014	6.4	4.0	2.9	6.0
742	Other nervous system	2,461	219	222	2,902	8.6	8.4	8.5	8.6
742.0	Encephalocele	350	17	26	393	1.2	0.7	1.0	1.2
742.1	Microcephalus	520	42	29	591	1.8	1.6	1.1	1.8
742.2	Brain reduction	553	55	58	666	1.9	2.1	2.2	2.0
742.3	Hydrocephalus	1,126	100	96	1,322	4.0	3.8	3.7	3.9
742.4-9	Other	209	29	35	273	0.7	1.1	1.3	0.8
743	Eye	855	78	76	1,009	3.0	3.0	2.9	3.0
743.0	Anophthalmos	82	7	9	98	0.3	0.3	0.3	0.3
743.1	Microphthalmos	264	21	25	310	0.9	0.8	1.0	0.9
743.2	Buphthalmos	64	4	4	72	0.2	0.2	0.2	0.2
743.3	Cataract & lens	259	24	14	297	0.9	0.9	0.5	0.9
743.32	Cataract	252	24	13	289	0.9	0.9	0.5	0.9
743.4-9	Other	309	28	34	371	1.1	1.1	1.3	1.1
744	Ear, face & neck	313	44	48	405	1.1	1.7	1.8	1.2
744.0	Ear-affecting hearing	240	30	21	291	0.8	1.2	0.8	0.9
744.00	Auditory canal a/s	206	23	16	245	0.7	0.9	0.6	0.7
744.01	Absent auricle	33	5	6	44	0.1	0.2	0.2	0.1
744.1-3	Other ear	35	4	19	58	0.1	0.2	0.7	0.2
744.4-9	Face & neck	48	12	8	68	0.2	0.5	0.3	0.2
745	Bulbus cordis & cardiac septal closure	6,457	717	736	7,910	22.7	27.5	28.2	23.5
745.0	Common truncus	203	23	7	233	0.7	0.9	0.3	0.7
745.1	Transpstn. grt. vessels	1,018	89	99	1,206	3.6	3.4	3.8	3.6
745.2	Tetralogy of Fallot	432	50	65	547	1.5	1.9	2.5	1.6
745.3	Common ventricle	187	14	12	213	0.7	0.5	0.5	0.6
745.4	Vent. septal defect	4,308	497	481	5,286	15.1	19.1	18.4	15.7
745.5	Ostium secundum ASD	1,219	170	154	1,543	4.3	6.5	5.9	4.6
745.6	Endocardial cushion	517	47	64	628	1.8	1.8	2.4	1.9
745.7-9	Other	36	4	5	45	0.1	0.2	0.2	0.1
746	Other heart	2,856	310	320	3,486	10.0	11.9	12.2	10.4
746.0	Pulmonary valve	1,062	127	108	1,297	3.7	4.9	4.1	3.9
746.00	atresia	377	33	27	437	1.3	1.3	1.0	1.3
746.01	stenosis	573	77	67	717	2.0	3.0	2.6	2.1
746.1	Tricuspid a/s	383	32	44	459	1.3	1.2	1.7	1.4
746.2	Ebstein anomaly	90	11	13	114	0.3	0.4	0.5	0.3
746.3-4	Aortic valve stenosis, insufficiency	332	38	46	416	1.2	1.5	1.8	1.2
746.5-6	Mitral st/insuffic.	211	24	26	261	0.7	0.9	1.0	0.8
746.7	Hypoplastic L heart	671	60	43	774	2.4	2.3	1.6	2.3
746.8	Other specified	428	48	68	544	1.5	1.8	2.6	1.6
746.9	Unspecified	202	29	30	261	0.7	1.1	1.1	0.8

Table 2.5: *Selected congenital malformations, Australia, 1981-1994 (cont.)*

Codes	Malformations	1981-1992	1993	1994	1981-94	1981-1992	1993	1994	1981-94
			Number				Rate per 10,000 births		
747	Circulatory	3,121	318	286	3,725	11.0	12.2	10.9	11.1
747.0	Pat. ductus arteriosus	1,928	211	176	2,315	6.8	8.1	6.7	6.9
747.1	Coarctation of aorta	839	73	61	973	2.9	2.8	2.3	2.9
747.2	Other aorta	336	25	28	389	1.2	1.0	1.1	1.2
747.3	Pulmonary artery	253	19	33	305	0.9	0.7	1.3	0.9
747.4	Great veins	315	32	27	374	1.1	1.2	1.0	1.1
747.42	Total anomalous pulm. venous return	186	20	15	221	0.7	0.8	0.6	0.7
747.6	Peripheral vascular	111	12	8	131	0.4	0.5	0.3	0.4
747.8	Other specified	33	4	3	40	0.1	0.2	0.1	0.1
747.9	Unspecified	2	3	3	8	0.0	0.1	0.1	0.0
748	Respiratory	824	78	58	960	2.9	3.0	2.2	2.9
748.0	Choanal atresia	287	30	26	343	1.0	1.2	1.0	1.0
748.1	Other nose	83	11	5	99	0.3	0.4	0.2	0.3
748.2-3	Larynx/ trach./ bronch.	168	14	8	190	0.6	0.5	0.3	0.6
748.4-6	Lung	301	25	19	345	1.1	1.0	0.7	1.0
748.8-9	Other respiratory	20	-	3	23	0.1	-	0.1	0.1
749	Cleft palate/ lip	4,196	402	382	4,980	14.7	15.4	14.6	14.8
749.0	Cleft palate	1,606	161	142	1,909	5.6	6.2	5.4	5.7
749.1	Cleft lip	885	87	80	1,052	3.1	3.3	3.1	3.1
749.2	Cleft palate + lip	1,703	154	160	2,017	6.0	5.9	6.1	6.0
750	Upper alimentary tract	930	104	70	1,104	3.3	4.0	2.7	3.3
750.3	TOF, oesophageal a/s	877	98	66	1,041	3.1	3.8	2.5	3.1
750.*	Other	58	8	6	72	0.2	0.3	0.2	0.2
751	Other digestive	2,517	217	208	2,942	8.8	8.3	8.0	8.7
751.1	Small intestine a/s	610	53	48	711	2.1	2.0	1.8	2.1
751.10	Duodenum a/s	370	35	31	436	1.3	1.3	1.2	1.3
751.11	Jejunum a/s	118	11	6	135	0.4	0.4	0.2	0.4
751.12	Ileum a/s	90	5	4	99	0.3	0.2	0.2	0.3
751.19	Unspecified a/s	50	2	7	59	0.2	0.1	0.3	0.2
751.2	Large intestine, rectum, anal canal a/s	961	85	77	1,123	3.4	3.3	2.9	3.3
751.20	Large intestine a/s	75	7	4	86	0.3	0.3	0.2	0.3
751.21-2	Rectum a/s	102	7	7	116	0.4	0.3	0.3	0.3
751.23-4	Anus a/s	833	79	66	978	2.9	3.0	2.5	2.9
751.3	Hirschsprung dis., etc	332	37	31	400	1.2	1.4	1.2	1.2
751.4	Intestinal fixation	367	14	20	401	1.3	0.5	0.8	1.2
751.5-9	Other digestive	435	43	44	522	1.5	1.7	1.7	1.6

Table 2.5: *Selected congenital malformations, Australia, 1981-1994 (cont.)*

Codes	Malformations	1981-1992	1993	1994	1981-94	1981-1992	1993	1994	1981-94
			Number			Rate per 10,000 births			
752	Genital organs	6,892	664	626	8,182	24.2	25.5	24.0	24.3
752.0-1	Ovaries/fallopian, etc	92	6	3	101	0.3	0.2	0.1	0.3
752.2-3	Uterus	169	11	9	189	0.6	0.4	0.3	0.6
752.4	Cerv., vagina, external	160	15	4	179	0.6	0.6	0.2	0.5
752.6	Hypospadias etc	5,911	571	550	7,032	20.8	21.9	21.0	20.9
752.60,3-5	Hypospadias	5,512	550	537	6,599	19.4	21.1	20.5	19.6
752.61	Epispadias	74	5	6	85	0.3	0.2	0.2	0.3
752.62	Chordee	895	61	25	981	3.1	2.3	1.0	2.9
752.7	Indeterminate sex, etc	426	42	47	515	1.5	1.6	1.8	1.5
752.74	Ambiguous genitalia	202	17	21	240	0.7	0.7	0.8	0.7
752.79	Indeterminate sex NOS	190	19	19	228	0.7	0.7	0.7	0.7
752.8	Other specified	375	40	29	444	1.3	1.5	1.1	1.3
752.9	Unspecified	21	-	1	22	0.1	-	0.0	0.1
753	Urinary	3,667	433	514	4,614	12.9	16.6	19.7	13.7
753.0	Renal agenesis/dysgen.	1,022	73	84	1,179	3.6	2.8	3.2	3.5
753.00	Bilateral	617	33	32	682	2.2	1.3	1.2	2.0
753.01	Unilateral	365	39	45	449	1.3	1.5	1.7	1.3
753.1	Cystic kidney disease	730	74	79	883	2.6	2.8	3.0	2.6
753.11-3	Polycystic	326	18	21	365	1.1	0.7	0.8	1.1
753.16	Multicystic	357	47	49	453	1.3	1.8	1.9	1.3
753.2	Obstructive defects renal pelvis/ureter	1,368	201	275	1,844	4.8	7.7	10.5	5.5
753.20	Hydronephrosis	884	130	157	1,171	3.1	5.0	6.0	3.5
753.21-9	Other	584	79	132	795	2.1	3.0	5.1	2.4
753.3	Other spec. kidney	439	56	57	552	1.5	2.1	2.2	1.6
753.32	Horseshoe kidney, etc	233	21	23	277	0.8	0.8	0.9	0.8
753.4	Other spec. ureter	164	27	34	225	0.6	1.0	1.3	0.7
753.5	Exstrophy of urinary bladder	100	8	6	114	0.4	0.3	0.2	0.3
753.6	Atresia/ stenosis of urethra, bladder neck	241	21	31	293	0.8	0.8	1.2	0.9
753.7	Urachus	31	1	-	32	0.1	0.0	-	0.1
753.8	Other bladder/ urethra	134	8	13	155	0.5	0.3	0.5	0.5
753.9	Unspecified	33	8	3	44	0.1	0.3	0.1	0.1
754	Certain musculoskeletal	6,324	562	484	7,370	22.2	21.6	18.5	21.9
754.30	Dislocation of hip	6,179	549	474	7,202	21.7	21.1	18.1	21.4
754.*	Other	153	13	10	176	0.5	0.5	0.4	0.5
755	Limb	3,938	418	379	4,735	13.8	16.0	14.5	14.1
755.0	Polydactyly	1,771	194	173	2,138	6.2	7.4	6.6	6.3
755.1	Syndactyly	704	64	73	841	2.5	2.5	2.8	2.5
755.2	Reduction, upper limb	978	89	73	1,140	3.4	3.4	2.8	3.4
755.3	Reduction, lower limb	460	34	37	531	1.6	1.3	1.4	1.6
755.4	Reduction, unspec limb	21	1	-	22	0.1	0.0	-	0.1
755.5	Other upper limb	289	34	36	359	1.0	1.3	1.4	1.1
755.6	Other lower limb	179	27	43	249	0.6	1.0	1.6	0.7
755.8	Other specified	202	26	16	244	0.7	1.0	0.6	0.7
755.80	Arthrogryposis multiplex congenita	159	20	15	194	0.6	0.8	0.6	0.6
755.9	Unspecified	8	2	2	12	0.0	0.1	0.1	0.0

Table 2.5: *Selected congenital malformations, Australia, 1981-1994 (cont.)*

Codes	Malformations	1981-1992	1993	1994	1981-94	1981-1992	1993	1994	1981-94
		Number				Rate per 10,000 births			
756	Other musculoskeletal	3,879	353	326	4,558	13.6	13.5	12.5	13.5
756.0	Skull, face & bones	847	77	73	997	3.0	3.0	2.8	3.0
756.00	Craniosynostosis	411	29	35	475	1.4	1.1	1.3	1.4
756.03	Pierre Robin synd.	260	28	21	309	0.9	1.1	0.8	0.9
756.1	Spine	499	53	53	605	1.8	2.0	2.0	1.8
756.3	Ribs & sternum	207	30	21	258	0.7	1.2	0.8	0.8
756.4	Chondrodystrophy	302	15	27	344	1.1	0.6	1.0	1.0
756.43	Achondroplasia	151	8	7	166	0.5	0.3	0.3	0.5
756.44	Other dwarfing synd.	104	4	9	117	0.4	0.2	0.3	0.3
756.5	Osteodystrophies	176	25	14	215	0.6	1.0	0.5	0.6
756.50	Osteogenesis imperfecta	135	11	9	155	0.5	0.4	0.3	0.5
756.6	Diaphragm	942	61	72	1,075	3.3	2.3	2.8	3.2
756.61	Diaphragmatic hernia	835	54	61	950	2.9	2.1	2.3	2.8
756.7	Abdominal wall	1,090	111	77	1,278	3.8	4.3	2.9	3.8
756.70	Exomphalos	636	47	30	713	2.2	1.8	1.1	2.1
756.71	Gastroschisis	324	53	40	417	1.1	2.0	1.5	1.2
756.8	Other specified	87	4	7	98	0.3	0.2	0.3	0.3
756.9	Unspecified	12	3	3	18	0.0	0.1	0.1	0.1
757	Integument	311	35	66	412	1.1	1.3	2.5	1.2
757.80	Cystic hygroma	209	22	22	253	0.7	0.8	0.8	0.8
758	Chromosomal	5,449	636	604	6,689	19.2	24.4	23.1	19.9
758.0	Trisomy 21 (Down)	3,422	366	320	4,108	12.0	14.0	12.2	12.2
758.1	Trisomy 13 (Patau)	263	28	22	313	0.9	1.1	0.8	0.9
758.2	Trisomy 18 (Edwards)	578	71	54	703	2.0	2.7	2.1	2.1
758.3	Autosomal deletion	211	24	45	280	0.7	0.9	1.7	0.8
758.5	Other autosomal	465	63	85	613	1.6	2.4	3.3	1.8
758.6	Turner syndrome	248	31	39	318	0.9	1.2	1.5	0.9
758.7	Klinefelter syndrome	84	18	11	113	0.3	0.7	0.4	0.3
758.8	Other sex chromosomes	174	34	26	234	0.6	1.3	1.0	0.7
758.9	Unspecified	24	2	5	31	0.1	0.1	0.2	0.1
759	Other & unspecified	1,125	129	104	1,358	4.0	5.0	4.0	4.0
759.0	Spleen	88	12	14	114	0.3	0.5	0.5	0.3
759.1	Adrenal gland	56	2	1	59	0.2	0.1	0.0	0.2
759.2	Other endocrine glands	109	10	11	130	0.4	0.4	0.4	0.4
759.3	Situs inversus	83	21	10	114	0.3	0.8	0.4	0.3
759.4	Conjoined twins	26	3	1	30	0.1	0.1	0.0	0.1
759.6	Hamartoses NEC	14	2	1	17	0.0	0.1	0.0	0.1
759.7	Multiple, so described	72	3	6	81	0.3	0.1	0.2	0.2
759.8	Other specified	729	83	64	876	2.6	3.2	2.4	2.6
759.9	Unspecified	12	3	1	16	0.0	0.1	0.0	0.0
760.2	Congenital rubella	19	2	2	23	0.1	0.1	0.1	0.1
760.70	Fetal hydantoin synd.	11	-	-	11	0.0	-	-	0.0
760.76	Fetal alcohol synd.	23	-	-	23	0.1	-	-	0.1

Note: Data for 1981 exclude Vic, WA, ACT, NT and certain hospital in NSW; data for 1982-1985 exclude NT.

Table 2.6: *Selected congenital malformations, by State or Territory of birth, 1990-1994*

Codes	Malformations	NSW	Vic	Qld	WA	SA	Tas	ACT	NT	Australia
						Number				
	Total									
	1993	1,261	1,379	901	350	379	73	41	42	4,426
	1994	1,104	1,565	910	228	350	60	39	41	4,297
	1990-94	6,861	7,072	4,380	1,564	1,836	392	236	223	22,564
740	Anencephalus									
	1993	16	24	12	10	1	-	-	1	64
	1994	12	11	10	6	1	-	1	3	44
	1990-94	71	66	73	45	8	6	4	10	283
741	Spina bifida									
	1993	44	22	20	6	6	4	-	1	103
	1994	17	22	17	14	4	1	2	-	77
	1990-94	196	149	126	74	32	13	7	8	605
742.0	Encephalocele									
	1993	10	1	1	2	2	-	1	-	17
	1994	2	14	3	4	2	1	-	-	26
	1990-94	35	42	11	14	7	1	2	1	113
742.1	Microcephalus									
	1993	15	10	9	5	2	-	-	1	42
	1994	4	13	3	3	4	-	-	2	29
	1990-94	70	58	29	20	10	2	2	6	197
742.3	Hydrocephalus									
	1993	22	36	16	9	8	6	-	3	100
	1994	21	38	15	11	5	1	3	2	96
	1990-94	162	157	75	42	30	15	14	10	505
745.1	Transposition of great vessels									
	1993	26	25	16	6	10	2	-	4	89
	1994	18	41	22	7	8	2	-	1	99
	1990-94	134	154	79	39	48	12	-	5	471
745.2	Tetralogy of Fallot									
	1993	16	18	9	4	-	-	-	3	50
	1994	17	31	4	7	3	-	1	2	65
	1990-94	94	107	37	24	14	-	5	6	287
745.4	Ventricular septal defect									
	1993	126	152	99	60	42	9	2	7	497
	1994	76	203	107	32	47	4	5	7	481
	1990-94	701	761	422	200	224	26	16	41	2,391
745.5	Ostium secundum atrial septal defect									
	1993	48	64	21	23	10	1	2	1	170
	1994	33	69	25	11	15	-	-	1	154
	1990-94	236	245	76	76	80	5	4	9	731

Table 2.6: *Selected congenital malformations, by State or Territory of birth, 1990-1994 (cont.)*

Codes	Malformations	NSW	Vic	Qld	WA	SA	Tas	ACT	NT	Australia
						Number				
746.00	Pulmonary valve atresia									
	1993	10	9	6	8	-	-	-	-	33
	1994	1	6	7	6	5	1	1	-	27
	1990-94	51	40	27	27	22	4	1	-	172
746.01	Pulmonary valve stenosis									
	1993	19	34	11	4	6	3	-	-	77
	1994	15	35	9	2	4	-	2	-	67
	1990-94	110	140	44	19	28	8	2	4	355
746.7	Hypoplastic left heart									
	1993	19	14	9	10	6	-	1	1	60
	1994	10	14	8	4	7	-	-	-	43
	1990-94	77	80	50	37	30	5	2	6	287
747.0	Patent ductus arteriosus									
	1993	86	69	15	23	14	1	2	1	211
	1994	54	73	18	22	7	-	1	1	176
	1990-94	427	327	86	100	57	6	8	2	1,013
747.1	Coarctation of aorta									
	1993	18	33	6	8	5	2	-	1	73
	1994	14	21	13	6	6	-	-	1	61
	1990-94	105	150	50	35	24	3	1	2	370
748.0	Choanal atresia									
	1993	9	15	2	3	1	-	-	-	30
	1994	10	10	3	-	1	1	1	-	26
	1990-94	42	64	15	9	13	4	4	1	152
749.0	Cleft palate									
	1993	60	37	33	16	8	4	3	-	161
	1994	51	45	19	11	11	3	2	-	142
	1990-94	282	196	128	73	61	19	8	10	777
749.1	Cleft lip									
	1993	25	29	14	6	11	1	-	1	87
	1994	25	22	18	8	4	2	1	-	80
	1990-94	142	136	75	22	31	9	7	3	425
749.2	Cleft palate + lip									
	1993	57	41	22	20	11	1	-	2	154
	1994	51	46	32	10	11	6	1	3	160
	1990-94	244	188	139	66	59	24	8	13	741
750.3	TOF, oesophageal atresia/stenosis									
	1993	32	23	20	5	13	3	-	2	98
	1994	16	24	11	5	8	2	-	-	66
	1990-94	130	114	73	26	50	12	3	3	411

Table 2.6: *Selected congenital malformations, by State or Territory of birth, 1990-1994 (cont.)*

Codes	Malformations	NSW	Vic	Qld	WA	SA	Tas	ACT	NT	Australia
						Number				
751.1	Small intestine atresia/stenosis									
	1993	11	11	12	6	10	3	-	-	53
	1994	15	11	16	2	3	1	-	-	48
	1990-94	83	77	50	28	33	4	2	2	279
751.2	Large intestine, rectum, anal canal atresia/stenosis									
	1993	24	24	16	9	9	-	-	3	85
	1994	18	26	23	6	3	1	-	-	77
	1990-94	127	128	72	42	42	1	1	7	420
751.3	Hirschsprung disease, etc									
	1993	6	18	6	4	3	-	-	-	37
	1994	2	15	4	6	4	-	-	-	31
	1990-94	42	79	22	21	12	1	1	-	178
752.60,	Hypospadias									
752.63-5	1993	154	170	109	57	48	2	7	3	550
	1994	164	185	94	24	52	10	6	2	537
	1990-94	901	908	478	235	237	48	34	6	2,847
752.7	Indeterminate sex, etc									
	1993	11	17	5	6	2	-	-	1	42
	1994	9	16	9	7	3	2	-	1	47
	1990-94	50	86	27	24	9	3	-	5	204
753.0	Renal agenesis/dysgenesis									
	1993	19	20	22	4	6	1	1	-	73
	1994	13	33	20	6	9	-	1	2	84
	1990-94	118	124	93	36	47	6	2	6	432
753.1	Cystic kidney disease									
	1993	21	22	11	11	5	1	-	3	74
	1994	23	24	18	3	8	-	1	2	79
	1990-94	109	121	69	35	45	5	2	12	398
753.2	Obstructive defects renal pelvis/ureter									
	1993	60	81	24	11	17	3	2	3	201
	1994	59	145	34	4	23	1	5	4	275
	1990-94	290	452	139	49	80	8	20	17	1,055
754.30	Dislocation of hip									
	1993	82	168	187	32	65	7	-	8	549
	1994	66	154	191	9	47	2	1	4	474
	1990-94	517	883	1,001	112	320	22	1	41	2,897

Table 2.6: *Selected congenital malformations, by State or Territory of birth, 1990-1994 (cont.)*

Codes	Malformations	NSW	Vic	Qld	WA	SA	Tas	ACT	NT	Australia
						Number				
755.0	Polydactyly									
	1993	77	59	32	4	18	2	2	-	194
	1994	67	51	32	8	12	2	1	-	173
	1990-94	350	278	156	56	72	10	5	-	927
755.1	Syndactyly									
	1993	24	17	14	4	2	3	-	-	64
	1994	19	25	15	3	9	-	2	-	73
	1990-94	107	105	70	20	38	4	3	1	348
755.2-4	Limb reduction									
	1993	39	26	33	10	8	2	-	2	120
	1994	41	22	19	6	9	2	-	2	101
	1990-94	220	131	119	43	66	10	5	8	602
756.00	Craniosynostosis									
	1993	14	8	4	1	2	-	-	-	29
	1994	3	24	5	1	2	-	-	-	35
	1990-94	152	67	14	8	18	1	1	2	263
756.4	Chondrodystrophy									
	1993	3	4	4	3	1	-	-	-	15
	1994	10	7	3	4	3	-	-	-	27
	1990-94	46	27	20	16	11	1	4	1	126
756.5	Osteodystrophies									
	1993	5	10	5	1	3	1	-	-	25
	1994	3	9	-	-	2	-	-	-	14
	1990-94	22	40	13	3	7	3	1	-	89
756.61	Diaphragmatic hernia									
	1993	11	15	17	4	6	-	1	-	54
	1994	19	19	16	3	2	1	1	-	61
	1990-94	108	112	86	30	28	11	4	1	380
756.70	Exomphalos									
	1993	12	12	7	7	6	2	-	1	47
	1994	12	5	5	5	2	1	-	-	30
	1990-94	77	70	38	31	26	6	3	1	252
756.71	Gastroschisis									
	1993	14	10	9	8	7	2	3	-	53
	1994	13	7	15	3	1	-	-	1	40
	1990-94	63	41	43	27	16	5	3	1	199

Table 2.6: *Selected congenital malformations, by State or Territory of birth, 1990-1994 (cont.)*

Codes	Malformations	NSW	Vic	Qld	WA	SA	Tas	ACT	NT	Australia
						Number				
758.0	Trisomy 21 (Down)									
	1993	149	76	64	38	22	5	8	4	366
	1994	117	88	62	18	18	7	7	3	320
	1990-94	600	446	303	142	104	42	33	17	1,687
758.1	Trisomy 13 (Patau)									
	1993	6	6	7	3	4	1	1	-	28
	1994	7	4	2	2	3	2	2	-	22
	1990-94	31	32	25	12	8	8	6	1	123
758.2	Trisomy 18 (Edwards)									
	1993	39	15	10	2	2	1	-	2	71
	1994	16	16	9	6	4	3	-	-	54
	1990-94	121	86	51	24	16	7	6	5	316
758.6	Turner syndrome									
	1993	16	8	2	3	1	1	-	-	31
	1994	19	6	8	2	3	-	-	1	39
	1990-94	66	41	23	13	10	2	1	2	158
758.3-5,	Other chromosomal									
758.7-9	1993	60	43	16	6	9	6	1	-	141
	1994	55	58	27	12	16	3	1	-	172
	1990-94	221	220	83	40	49	17	13	3	646

Table 2.7: *Selected congenital malformation rates, by State or Territory of birth, 1990-1994*

Codes	Malformations	NSW	Vic	Qld	WA	SA	Tas	ACT	NT	Australia
					Rate per 10,000 births					
	Total									
	1993	143.5	213.0	189.9	138.1	189.8	106.5	85.6	118.5	169.9
	1994	125.5	241.0	189.4	89.7	176.8	87.8	81.5	116.2	164.4
	1990-94	155.0	215.1	189.1	123.6	184.0	112.9	99.5	125.5	172.8
740	Anencephalus									
	1993	1.8	3.7	2.5	3.9	0.5	-	-	2.8	2.5
	1994	1.4	1.7	2.1	2.4	0.5	-	2.1	8.5	1.7
	1990-94	1.6	2.0	3.2	3.6	0.8	1.7	1.7	5.6	2.2
741	Spina bifida									
	1993	5.0	3.4	4.2	2.4	3.0	5.8	-	2.8	4.0
	1994	1.9	3.4	3.5	5.5	2.0	1.5	4.2	-	2.9
	1990-94	4.4	4.5	5.4	5.8	3.2	3.7	3.0	4.5	4.6
742.0	Encephalocele									
	1993	1.1	0.2	0.2	0.8	1.0	-	2.1	-	0.7
	1994	0.2	2.2	0.6	1.6	1.0	1.5	-	-	1.0
	1990-94	0.8	1.3	0.5	1.1	0.7	0.3	0.8	0.6	0.9
742.1	Microcephalus									
	1993	1.7	1.5	1.9	2.0	1.0	-	-	2.8	1.6
	1994	0.5	2.0	0.6	1.2	2.0	-	-	5.7	1.1
	1990-94	1.6	1.8	1.3	1.6	1.0	0.6	0.8	3.4	1.5
742.3	Hydrocephalus									
	1993	2.5	5.6	3.4	3.6	4.0	8.8	-	8.5	3.8
	1994	2.4	5.9	3.1	4.3	2.5	1.5	6.3	5.7	3.7
	1990-94	3.7	4.8	3.2	3.3	3.0	4.3	5.9	5.6	3.9
745.1	Transposition of great vessels									
	1993	3.0	3.9	3.4	2.4	5.0	2.9	-	11.3	3.4
	1994	2.0	6.3	4.6	2.8	4.0	2.9	-	2.8	3.8
	1990-94	3.0	4.7	3.4	3.1	4.8	3.5	-	2.8	3.6
745.2	Tetralogy of Fallot									
	1993	1.8	2.8	1.9	1.6	-	-	-	8.5	1.9
	1994	1.9	4.8	0.8	2.8	1.5	-	2.1	5.7	2.5
	1990-94	2.1	3.3	1.6	1.9	1.4	-	2.1	3.4	2.2
745.4	Ventricular septal defect									
	1993	14.3	23.5	20.9	23.7	21.0	13.1	4.2	19.7	19.1
	1994	8.6	31.3	22.3	12.6	23.7	5.9	10.5	19.8	18.4
	1990-94	15.8	23.2	18.2	15.8	22.5	7.5	6.7	23.1	18.3
745.5	Ostium secundum atrial septal defect									
	1993	5.5	9.9	4.4	9.1	5.0	1.5	4.2	2.8	6.5
	1994	3.8	10.6	5.2	4.3	7.6	-	-	2.8	5.9
	1990-94	5.3	7.5	3.3	6.0	8.0	1.4	1.7	5.1	5.6

Table 2.7: *Selected congenital malformation rates, by State or Territory of birth, 1990-1994 (cont.)*

Codes	Malformations	NSW	Vic	Qld	WA	SA	Tas	ACT	NT	Australia
					Rate per 10,000 births					
746.00	Pulmonary valve atresia									
	1993	1.1	1.4	1.3	3.2	-	-	-	-	1.3
	1994	0.1	0.9	1.5	2.4	2.5	1.5	2.1	-	1.0
	1990-94	1.2	1.2	1.2	2.1	2.2	1.2	0.4	-	1.3
746.01	Pulmonary valve stenosis									
	1993	2.2	5.3	2.3	1.6	3.0	4.4	-	-	3.0
	1994	1.7	5.4	1.9	0.8	2.0	-	4.2	-	2.6
	1990-94	2.5	4.3	1.9	1.5	2.8	2.3	0.8	2.3	2.7
746.7	Hypoplastic left heart									
	1993	2.2	2.2	1.9	3.9	3.0	-	2.1	2.8	2.3
	1994	1.1	2.2	1.7	1.6	3.5	-	-	-	1.6
	1990-94	1.7	2.4	2.2	2.9	3.0	1.4	0.8	3.4	2.2
747.0	Patent ductus arteriosus									
	1993	9.8	10.7	3.2	9.1	7.0	1.5	4.2	2.8	8.1
	1994	6.1	11.2	3.7	8.7	3.5	-	2.1	2.8	6.7
	1990-94	9.6	9.9	3.7	7.9	5.7	1.7	3.4	1.1	7.8
747.1	Coarctation of aorta									
	1993	2.0	5.1	1.3	3.2	2.5	2.9	-	2.8	2.8
	1994	1.6	3.2	2.7	2.4	3.0	-	-	2.8	2.3
	1990-94	2.4	4.6	2.2	2.8	2.4	0.9	0.4	1.1	2.8
748.0	Choanal atresia									
	1993	1.0	2.3	0.4	1.2	0.5	-	-	-	1.2
	1994	1.1	1.5	0.6	-	0.5	1.5	2.1	-	1.0
	1990-94	0.9	1.9	0.6	0.7	1.3	1.2	1.7	0.6	1.2
749.0	Cleft palate									
	1993	6.8	5.7	7.0	6.3	4.0	5.8	6.3	-	6.2
	1994	5.8	6.9	4.0	4.3	5.6	4.4	4.2	-	5.4
	1990-94	6.4	6.0	5.5	5.8	6.1	5.5	3.4	5.6	6.0
749.1	Cleft lip									
	1993	2.8	4.5	3.0	2.4	5.5	1.5	-	2.8	3.3
	1994	2.8	3.4	3.7	3.1	2.0	2.9	2.1	-	3.1
	1990-94	3.2	4.1	3.2	1.7	3.1	2.6	3.0	1.7	3.3
749.2	Cleft palate + lip									
	1993	6.5	6.3	4.6	7.9	5.5	1.5	-	5.6	5.9
	1994	5.8	7.1	6.7	3.9	5.6	8.8	2.1	8.5	6.1
	1990-94	5.5	5.7	6.0	5.2	5.9	6.9	3.4	7.3	5.7
750.3	TOF, oesophageal atresia/stenosis									
	1993	3.6	3.6	4.2	2.0	6.5	4.4	-	5.6	3.8
	1994	1.8	3.7	2.3	2.0	4.0	2.9	-	-	2.5
	1990-94	2.9	3.5	3.2	2.1	5.0	3.5	1.3	1.7	3.1

Table 2.7: *Selected congenital malformation rates, by State or Territory of birth, 1990-1994 (cont.)*

Codes	Malformations	NSW	Vic	Qld	WA	SA	Tas	ACT	NT	Australia
					Rate per 10,000 births					
751.1	Small intestine atresia/stenosis									
	1993	1.3	1.7	2.5	2.4	5.0	4.4	-	-	2.0
	1994	1.7	1.7	3.3	0.8	1.5	1.5	-	-	1.8
	1990-94	1.9	2.3	2.2	2.2	3.3	1.2	0.8	1.1	2.1
751.2	Large intestine, rectum, anal canal atresia/stenosis									
	1993	2.7	3.7	3.4	3.6	4.5	-	-	8.5	3.3
	1994	2.0	4.0	4.8	2.4	1.5	1.5	-	-	2.9
	1990-94	2.9	3.9	3.1	3.3	4.2	0.3	0.4	3.9	3.2
751.3	Hirschsprung disease, etc									
	1993	0.7	2.8	1.3	1.6	1.5	-	-	-	1.4
	1994	0.2	2.3	0.8	2.4	2.0	-	-	-	1.2
	1990-94	0.9	2.4	0.9	1.7	1.2	0.3	0.4	-	1.4
752.60,	Hypospadias									
752.63-5	1993	17.5	26.3	23.0	22.5	24.0	2.9	14.6	8.5	21.1
	1994	18.6	28.5	19.6	9.4	26.3	14.6	12.5	5.7	20.5
	1990-94	20.4	27.6	20.6	18.6	23.8	13.8	14.3	3.4	21.8
752.7	Indeterminate sex, etc									
	1993	1.3	2.6	1.1	2.4	1.0	-	-	2.8	1.6
	1994	1.0	2.5	1.9	2.8	1.5	2.9	-	2.8	1.8
	1990-94	1.1	2.6	1.2	1.9	0.9	0.9	-	2.8	1.6
753.0	Renal agenesis/dysgenesis									
	1993	2.2	3.1	4.6	1.6	3.0	1.5	2.1	-	2.8
	1994	1.5	5.1	4.2	2.4	4.5	-	2.1	5.7	3.2
	1990-94	2.7	3.8	4.0	2.8	4.7	1.7	0.8	3.4	3.3
753.1	Cystic kidney disease									
	1993	2.4	3.4	2.3	4.3	2.5	1.5	-	8.5	2.8
	1994	2.6	3.7	3.7	1.2	4.0	-	2.1	5.7	3.0
	1990-94	2.5	3.7	3.0	2.8	4.5	1.4	0.8	6.8	3.0
753.2	Obstructive defects renal pelvis/ureter									
	1993	6.8	12.5	5.1	4.3	8.5	4.4	4.2	8.5	7.7
	1994	6.7	22.3	7.1	1.6	11.6	1.5	10.5	11.3	10.5
	1990-94	6.6	13.8	6.0	3.9	8.0	2.3	8.4	9.6	8.1
754.30	Dislocation of hip									
	1993	9.3	26.0	39.4	12.6	32.6	10.2	-	22.6	21.1
	1994	7.5	23.7	39.8	3.5	23.7	2.9	2.1	11.3	18.1
	1990-94	11.7	26.9	43.2	8.9	32.1	6.3	0.4	23.1	22.2

Table 2.7: *Selected congenital malformation rates, by State or Territory of birth, 1990-1994 (cont.)*

Codes	Malformations	NSW	Vic	Qld	WA	SA	Tas	ACT	NT	Australia
					Rate per 10,000 births					
755.0	Polydactyly									
	1993	8.8	9.1	6.7	1.6	9.0	2.9	4.2	-	7.4
	1994	7.6	7.9	6.7	3.1	6.1	2.9	2.1	-	6.6
	1990-94	7.9	8.5	6.7	4.4	7.2	2.9	2.1	-	7.1
755.1	Syndactyly									
	1993	2.7	2.6	3.0	1.6	1.0	4.4	-	-	2.5
	1994	2.2	3.9	3.1	1.2	4.5	-	4.2	-	2.8
	1990-94	2.4	3.2	3.0	1.6	3.8	1.2	1.3	0.6	2.7
755.2-4	Limb reduction									
	1993	4.4	4.0	7.0	3.9	4.0	2.9	-	5.6	4.6
	1994	4.7	3.4	4.0	2.4	4.5	2.9	-	5.7	3.9
	1990-94	5.0	4.0	5.1	3.4	6.6	2.9	2.1	4.5	4.6
756.00	Craniosynostosis									
	1993	1.6	1.2	0.8	0.4	1.0	-	-	-	1.1
	1994	0.3	3.7	1.0	0.4	1.0	-	-	-	1.3
	1990-94	3.4	2.0	0.6	0.6	1.8	0.3	0.4	1.1	2.0
756.4	Chondrodystrophy									
	1993	0.3	0.6	0.8	1.2	0.5	-	-	-	0.6
	1994	1.1	1.1	0.6	1.6	1.5	-	-	-	1.0
	1990-94	1.0	0.8	0.9	1.3	1.1	0.3	1.7	0.6	1.0
756.5	Osteodystrophies									
	1993	0.6	1.5	1.1	0.4	1.5	1.5	-	-	1.0
	1994	0.3	1.4	-	-	1.0	-	-	-	0.5
	1990-94	0.5	1.2	0.6	0.2	0.7	0.9	0.4	-	0.7
756.61	Diaphragmatic hernia									
	1993	1.3	2.3	3.6	1.6	3.0	-	2.1	-	2.1
	1994	2.2	2.9	3.3	1.2	1.0	1.5	2.1	-	2.3
	1990-94	2.4	3.4	3.7	2.4	2.8	3.2	1.7	0.6	2.9
756.70	Exomphalos									
	1993	1.4	1.9	1.5	2.8	3.0	2.9	-	2.8	1.8
	1994	1.4	0.8	1.0	2.0	1.0	1.5	-	-	1.1
	1990-94	1.7	2.1	1.6	2.4	2.6	1.7	1.3	0.6	1.9
756.71	Gastroschisis									
	1993	1.6	1.5	1.9	3.2	3.5	2.9	6.3	-	2.0
	1994	1.5	1.1	3.1	1.2	0.5	-	-	2.8	1.5
	1990-94	1.4	1.2	1.9	2.1	1.6	1.4	1.3	0.6	1.5

Table 2.7: *Selected congenital malformation rates, by State or Territory of birth, 1990-1994 (cont.)*

Codes	Malformations	NSW	Vic	Qld	WA	SA	Tas	ACT	NT	Australia
					Rate per 10,000 births					
758.0	Trisomy 21 (Down)									
	1993	17.0	11.7	13.5	15.0	11.0	7.3	16.7	11.3	14.0
	1994	13.3	13.6	12.9	7.1	9.1	10.2	14.6	8.5	12.2
	1990-94	13.6	13.6	13.1	11.2	10.4	12.1	13.9	9.6	12.9
758.1	Trisomy 13 (Patau)									
	1993	0.7	0.9	1.5	1.2	2.0	1.5	2.1	-	1.1
	1994	0.8	0.6	0.4	0.8	1.5	2.9	4.2	-	0.8
	1990-94	0.7	1.0	1.1	0.9	0.8	2.3	2.5	0.6	0.9
758.2	Trisomy 18 (Edwards)									
	1993	4.4	2.3	2.1	0.8	1.0	1.5	-	5.6	2.7
	1994	1.8	2.5	1.9	2.4	2.0	4.4	-	-	2.1
	1990-94	2.7	2.6	2.2	1.9	1.6	2.0	2.5	2.8	2.4
758.6	Turner syndrome									
	1993	1.8	1.2	0.4	1.2	0.5	1.5	-	-	1.2
	1994	2.2	0.9	1.7	0.8	1.5	-	-	2.8	1.5
	1990-94	1.5	1.2	1.0	1.0	1.0	0.6	0.4	1.1	1.2
758.3-5,	Other chromosomal									
758.7-9	1993	6.8	6.6	3.4	2.4	4.5	8.8	2.1	-	5.4
	1994	6.3	8.9	5.6	4.7	8.1	4.4	2.1	-	6.6
	1990-94	5.0	6.7	3.6	3.2	4.9	4.9	5.5	1.7	4.9

Table 2.8: *Proportion of notified births with missing information, 1992-1994*

Characteristic	Births								
	NSW	Vic	Qld	WA	SA	Tas	ACT	NT	Australia
					Per cent				
Maternal age	2.8	1.1	1.8	2.1	-	7.9	8.5	0.7	1.9
Maternal country of birth	13.6	34.3	3.4	99.9	37.7	8.3	25.6	3.0	25.8
Maternal race	10.8	35.8	4.2	4.3	3.7	12.3	27.9	1.5	16.5
Plurality	1.7	0.7	0.8	0.2	0.2	3.5	-	0.7	1.0
Infant's sex	0.5	0.2	0.5	0.1	0.6	1.3	0.8	0.7	0.4
Birthweight	7.3	2.5	3.0	3.6	3.6	9.6	21.7	1.5	4.4
Gestational age	6.4	33.2	1.7	3.1	0.1	6.6	10.9	5.2	13.2

Table 2.9: *Terminations of pregnancy for fetal malformations, Australia, 1991-1994*

Congenital malformation	Induced abortions* (less than 20 weeks)				Induced births (20-27 weeks)				All terminations (up to 27 weeks)			
	1991	1992	1993	1994	1991	1992	1993	1994	1991	1992	1993	1994
						Number						
Anencephalus	52	62	56	84	10	9	24	10	62	71	80	94
Spina bifida	26	45	51	70	14	18	17	7	40	63	68	77
Encephalocele	8	6	9	15	-	3	3	2	8	9	12	17
Hydrocephalus	17	17	30	31	12	16	14	18	29	33	44	49
Cardiac defects	24	19	39	55	11	23	32	40	35	42	71	95
Renal abnormalities	3	6	15	18	10	3	7	11	13	9	22	29
Limb reduction defects	5	8	10	16	2	7	9	5	7	15	19	21
Diaphragmatic hernia	6	8	6	8	5	4	4	3	11	12	10	11
Exomphalos	12	13	23	24	5	8	6	1	17	21	29	25
Trisomy 21	94	113	118	128	10	20	20	25	104	133	138	153
Trisomy 13	13	7	22	16	2	4	6	7	15	11	28	23
Trisomy 18	29	40	34	48	6	11	9	9	35	51	43	57
Other autosomal	25	43	43	46	13	10	8	14	38	53	51	60
Gonadal dysgenesis	16	22	18	25	-	7	2	9	16	29	20	34
Sex chromosomal	19	11	16	22	1	5	2	2	20	16	18	24
Other	22	28	43	65	13	16	13	14	35	44	56	79
Single	94	130	135	210	40	49	62	56	134	179	197	266
Multiple	29	35	50	65	20	20	15	11	49	55	65	76
Syndrome	202	241	269	304	36	62	55	72	238	303	324	376
All fetuses	325	406	454	579	96	131	132	139	421	537	586	718
						Ratio per 10,000 births						
Anencephalus	2.0	2.4	2.1	3.2	0.4	0.3	0.9	0.4	2.4	2.7	3.1	3.6
Spina bifida	1.0	1.7	2.0	2.7	0.5	0.7	0.7	0.3	1.6	2.4	2.6	2.9
Encephalocele	0.3	0.2	0.3	0.6	-	0.1	0.1	0.1	0.3	0.3	0.5	0.7
Hydrocephalus	0.7	0.6	1.2	1.2	0.5	0.6	0.5	0.7	1.1	1.3	1.7	1.9
Cardiac defects	0.9	0.7	1.5	2.1	0.4	0.9	1.2	1.5	1.4	1.6	2.7	3.6
Renal abnormalities	0.1	0.2	0.6	0.7	0.4	0.1	0.3	0.4	0.5	0.3	0.8	1.1
Limb reduction defects	0.2	0.3	0.4	0.6	0.1	0.3	0.3	0.2	0.3	0.6	0.7	0.8
Diaphragmatic hernia	0.2	0.3	0.2	0.3	0.2	0.2	0.2	0.1	0.4	0.5	0.4	0.4
Exomphalos	0.5	0.5	0.9	0.9	0.2	0.3	0.2	0.0	0.7	0.8	1.1	1.0
Trisomy 21	3.7	4.3	4.5	4.9	0.4	0.8	0.8	1.0	4.1	5.1	5.3	5.9
Trisomy 13	0.5	0.3	0.8	0.6	0.1	0.2	0.2	0.3	0.6	0.4	1.1	0.9
Trisomy 18	1.1	1.5	1.3	1.8	0.2	0.4	0.3	0.3	1.4	1.9	1.7	2.2
Other autosomal	1.0	1.6	1.7	1.8	0.5	0.4	0.3	0.5	1.5	2.0	2.0	2.3
Gonadal dysgenesis	0.6	0.8	0.7	1.0	-	0.3	0.1	0.3	0.6	1.1	0.8	1.3
Sex chromosomal	0.7	0.4	0.6	0.8	0.0	0.2	0.1	0.1	0.8	0.6	0.7	0.9
Other	0.9	1.1	1.7	2.5	0.5	0.6	0.5	0.5	1.4	1.7	2.1	3.0
Single	3.7	4.9	5.2	8.0	1.6	1.9	2.4	2.1	5.2	6.8	7.6	10.2
Multiple	1.1	1.3	1.9	2.5	0.8	0.8	0.6	0.4	1.9	2.1	2.5	2.9
Syndrome	7.9	9.2	10.3	11.6	1.4	2.4	2.1	2.8	9.3	11.5	12.4	14.4
All fetuses	12.7	15.5	17.4	22.2	3.7	5.0	5.1	5.3	16.4	20.4	22.5	27.5

* *Includes terminations at unstated gestational ages*

Table 2.10 *Terminations of pregnancy for fetal malformations by gestational age, Australia, 1991-1994*

Years	Gestational age (weeks)										
	<10	**10**	**11**	**12**	**13**	**14**	**15**	**16**	**17**	**18**	**19**
1991	-	5	12	16	15	12	12	19	31	76	66
1992	3	5	5	22	18	16	14	34	51	94	80
1993	1	3	6	12	19	27	16	29	38	104	119
1994	4	5	6	16	15	14	22	32	47	103	101
All years	8	18	29	66	67	69	64	114	167	377	366

	20	**21**	**22**	**23**	**24**	**25**	**26**	**27**	**Not known**	**All terminations**
1991	33	16	14	13	6	4	5	4	62	421
1992	42	32	30	6	6	6	4	3	66	537
1993	42	27	27	8	10	10	4	3	81	586
1994	52	30	20	16	6	2	5	3	219	718
All years	169	105	91	43	28	22	18	13	428	2,262

Table 2.11 *Terminations of pregnancy for selected malformations by gestational age, Australia, 1991-1994*

Malformation	Gestational age (weeks)										
	<10	**10**	**11**	**12**	**13**	**14**	**15**	**16**	**17**	**18**	**19**
Anencephalus	-	2	-	2	3	15	8	24	41	66	49
Spina bifida	1	-	1	1	1	1	1	8	23	63	52
Trisomy 21	5	9	18	27	27	20	19	31	34	83	85
Other malformations	2	7	10	36	36	33	36	51	69	165	180
All malformations	8	18	29	66	67	69	64	114	167	377	366

	20	**21**	**22**	**23**	**24**	**25**	**26**	**27**	**Not known**	**All terminations**
Anencephalus	15	11	9	3	3	4	2	5	45	307
Spina bifida	22	12	9	4	3	4	2	-	40	248
Trisomy 21	37	15	13	4	5	1	-	-	95	528
Other malformations	95	67	60	32	17	13	14	8	248	1,179
All malformations	169	105	91	43	28	22	18	13	428	2,262

Table 2.12: *Terminations of pregnancy for fetal malformations, States and Territories, 1991-1994*

State / Territory	Induced abortions* (less than 20 weeks)				Induced births (20-27 weeks)				All terminations (up to 27 weeks)			
	1991	1992	1993	1994	1991	1992	1993	1994	1991	1992	1993	1994
	Number											
New South Wales	72	152	132	151	7	22	7	13	79	174	139	164
Victoria	123	112	149	225	44	63	72	63	167	175	221	288
Queensland	14	16	19	16	9	8	7	4	23	24	26	20
Western Australia	47	58	70	91	9	3	14	10	56	61	84	101
South Australia	55	59	59	58	23	26	25	36	78	85	84	94
Tasmania	4	5	12	20	1	-	1	2	5	5	13	22
Aust. Capital Territory	10	4	9	14	3	5	5	8	13	9	14	22
Northern Territory	-	-	4	4	-	4	1	3	-	4	5	7
Australia	325	406	454	579	96	131	132	139	421	537	586	718

* *Includes terminations of unstated gestational ages*

Table 2.13: *Ratios of terminations of pregnancy for fetal malformations, States and Territories, 1994*

State / Territory	Terminations of pregnancy			Total births	Ratio of TOPs per 10,000 births		
	<20	20+	All TOPs		<20	20+	All TOPs
	Number				Ratio		
New South Wales	151	13	164	87,984	17.2	1.5	18.6
Victoria	225	63	288	64,932	34.7	9.7	44.4
Queensland	16	4	20	48,046	3.3	0.8	4.2
Western Australia	91	10	101	25,425	35.8	3.9	39.7
South Australia	58	36	94	19,801	29.3	18.2	47.5
Tasmania	20	2	22	6,836	29.3	2.9	32.2
Australian Capital Territory	14	8	22	4,784	29.3	16.7	46.0
Northern Territory	4	3	7	3,527	11.3	8.5	19.8
Australia	579	139	718	261,335	22.2	5.3	27.5

TOPs: Terminations of pregnancy

Table 2.14 *Terminations of pregnancy for fetal malformations by State, Australia, 1991-1994*

State	Gestational age (weeks)										
	<10	**10**	**11**	**12**	**13**	**14**	**15**	**16**	**17**	**18**	**19**
New South Wales	6	7	10	22	10	20	25	39	52	101	81
Victoria	1	7	14	30	36	25	20	33	37	80	81
Queensland	1	1	-	1	1	1	1	4	8	16	14
Western Australia	-	1	-	5	3	10	8	25	33	68	105
South Australia	-	1	4	6	13	10	5	12	31	82	65
Other	-	1	1	2	4	3	5	1	6	30	20
Australia	8	18	29	66	67	69	64	114	167	377	366

	20	**21**	**22**	**23**	**24**	**25**	**26**	**27**	**Not known**	**All terminations**
New South Wales	21	9	5	3	2	2	2	-	139	556
Victoria	77	52	47	25	13	10	8	6	249	851
Queensland	13	3	6	1	1	1	1	2	17	93
Western Australia	11	3	7	5	3	4	1	2	-	302
South Australia	37	31	16	8	7	3	6	2	-	341
Other	10	7	10	1	2	2	-	1	13	119
Australia	169	105	91	43	28	22	18	13	418	2,262

Table 2.15: *Proportion of notified terminations of pregnancy with missing information, 1992-1994*

Characteristic	Terminations of pregnancy (<20 weeks)								
	NSW	**Vic**	**Qld**	**WA**	**SA**	**Tas**	**ACT**	**NT**	**Australia**
	Per cent								
Maternal age	6.0	1.9	3.9	2.7	0.6	-	-	-	3.1
Maternal country of birth	62.3	86.4	100.0	100.0	100.0	100.0	92.6	100.0	83.9
Maternal race	59.5	98.8	100.0	100.0	100.0	100.0	92.6	37.5	86.9
Plurality	5.3	1.6	7.8	1.4	2.8	27.0	-	75.0	4.1
Infant's sex	2.1	14.6	3.9	6.4	15.3	64.9	-	87.5	10.7
Birthweight	73.8	77.6	98.0	100.0	100.0	100.0	88.9	100.0	84.2
Gestational age	26.4	43.4	31.4	3.2	0.6	16.2	7.4	-	24.9

Table 2.16: *Congenital malformations by maternal age, Australia, 1992-1994*

Maternal age (years)	Type of malformation				
	Isolated	Multiple	Syndrome		All types
			Chromosomal	Non-chromosomal	
			Number		
Births (20+ weeks)					
Less than 20	566	62	40	36	704
20 - 24	1,961	177	182	83	2,403
25 - 29	3,371	255	364	132	4,122
30 - 34	2,924	224	496	124	3,768
35 - 39	1,074	88	383	40	1,585
40 years and over	174	20	188	12	394
Not stated	90	16	135	10	251
All ages	10,160	842	1,788	437	13,227
Terminations of pregnancy (<20 weeks)					
Less than 20	18	10	2	1	31
20 - 24	88	24	35	8	155
25 - 29	164	49	105	16	334
30 - 34	129	42	113	10	294
35 - 39	55	16	307	3	381
40 years and over	10	2	188	-	200
Not stated	11	7	24	2	44
All ages	475	150	774	40	1,439
			Rate per 10,000 births		
Births (20+ weeks)					
Less than 20	134.6	14.7	9.5	8.6	167.5
20 - 24	126.3	11.4	11.7	5.3	154.8
25 - 29	127.3	9.6	13.8	5.0	155.7
30 - 34	128.0	9.8	21.7	5.4	164.9
35 - 39	133.3	10.9	47.5	5.0	196.7
40 years and over	135.1	15.5	146.0	9.3	305.9
All ages	129.5	10.7	22.8	5.6	168.6
Terminations of pregnancy (<20 weeks)					
			Ratio per 10,000 births		
Less than 20	4.3	2.4	0.5	0.2	7.4
20 - 24	5.7	1.5	2.3	0.5	10.0
25 - 29	6.2	1.9	4.0	0.6	12.6
30 - 34	5.6	1.8	4.9	0.4	12.9
35 - 39	6.8	2.0	38.1	0.4	47.3
40 years and over	7.8	1.6	146.0	-	155.3
All ages	6.1	1.9	9.9	0.5	18.3

Table 2.17: *Congenital malformations by plurality, Australia, 1992-1994*

Plurality	Type of malformation				
	Isolated	Multiple	Syndrome		All types
			Chromosomal	Non-chromosomal	
			Number		
Births (20+ weeks)					
Singleton	9,716	779	1,665	412	12,572
Twins	387	55	43	20	505
Others	20	2	1	-	23
Not stated	37	6	79	5	127
All pluralities	10,160	842	1,788	437	13,227
Terminations of pregnancy (<20 weeks)					
Singleton	440	143	724	39	1,346
Twins	17	3	10	1	31
Others	-	-	3	-	3
Not stated	18	4	37	-	59
All pluralities	475	150	774	40	1,439
			Rate per 10,000 births		
Births (20+ weeks)					
Singleton	127.3	10.2	21.8	5.4	164.7
Twins	188.6	26.8	21.0	9.7	246.1
Others	220.8	22.1	11.0	-	253.9
All pluralities	129.5	10.7	22.8	5.6	168.6
Terminations of pregnancy (<20 weeks)					
			Ratio per 10,000 births		
Singleton	5.8	1.9	9.5	0.5	17.6
Twins	8.3	1.5	4.9	0.5	15.1
Others	-	-	33.1	-	33.1
All pluralities	6.1	1.9	9.9	0.5	18.3

Table 2.18: *Selected congenital malformations by maternal country of birth, Australia, 1991-1994*

Congenital malformation	Australia		New Zealand	United Kingdom	Other Europe	Lebanon
	Non-Indigenous	Indigenous				
Total births	640,816	21,770	20,412	35,946	32,636	12,700
			Number			
Anencephalus	109	10	4	3	8	4
Spina bifida	257	14	7	12	16	6
Encephalocele	41	5	-	1	4	2
Hydrocephalus	174	20	4	12	16	7
Transposition of great vessels	198	4	7	4	17	7
Ventricular septal defect	939	46	31	40	60	13
Hypoplastic left heart	122	6	1	4	9	4
Cleft palate	324	10	10	17	29	10
Cleft lip	518	22	9	25	30	12
Oesophageal atresia or stenosis	180	1	6	6	15	3
Small intestinal atresia or stenosis	107	3	6	5	7	6
Anorectal atresia or stenosis	168	6	2	8	11	3
Hypospadias	1,322	31	30	61	83	33
Renal agenesis and dysgenesis	183	10	4	6	11	5
Cystic kidney disease	165	7	1	10	12	4
Obstructive defects of renal pelvis and ureter	418	17	11	26	30	9
Congenital dislocation of hip	1,392	25	45	86	70	6
Limb reduction defects	280	12	8	15	17	9
Diaphragmatic hernia	167	4	6	13	12	4
Exomphalos	90	3	1	6	8	2
Gastroschisis	99	6	1	3	7	-
Trisomy 21	603	25	12	20	61	16
Trisomy 18	113	4	-	7	8	5
All infants and fetuses	9,982	352	270	519	683	229
			Rate per 10,000 births			
Anencephalus	1.7	4.6	2.0	0.8	2.5	3.1
Spina bifida	4.0	6.4	3.4	3.3	4.9	4.7
Encephalocele	0.6	2.3	-	0.3	1.2	1.6
Hydrocephalus	2.7	9.2	2.0	3.3	4.9	5.5
Transposition of great vessels	3.1	1.8	3.4	1.1	5.2	5.5
Ventricular septal defect	14.7	21.1	15.2	11.1	18.4	10.2
Hypoplastic left heart	1.9	2.8	0.5	1.1	2.8	3.1
Cleft palate	5.1	4.6	4.9	4.7	8.9	7.9
Cleft lip	8.1	10.1	4.4	7.0	9.2	9.4
Oesophageal atresia or stenosis	2.8	0.5	2.9	1.7	4.6	2.4
Small intestinal atresia or stenosis	1.7	1.4	2.9	1.4	2.1	4.7
Anorectal atresia or stenosis	2.6	2.8	1.0	2.2	3.4	2.4
Hypospadias	20.6	14.2	14.7	17.0	25.4	26.0
Renal agenesis and dysgenesis	2.9	4.6	2.0	1.7	3.4	3.9
Cystic kidney disease	2.6	3.2	0.5	2.8	3.7	3.1
Obstructive defects of renal pelvis and ureter	6.5	7.8	5.4	7.2	9.2	7.1
Congenital dislocation of hip	21.7	11.5	22.0	23.9	21.4	4.7
Limb reduction defects	4.4	5.5	3.9	4.2	5.2	7.1
Diaphragmatic hernia	2.6	1.8	2.9	3.6	3.7	3.1
Exomphalos	1.4	1.4	0.5	1.7	2.5	1.6
Gastroschisis	1.5	2.8	0.5	0.8	2.1	-
Trisomy 21	9.4	11.5	5.9	5.6	18.7	12.6
Trisomy 18	1.8	1.8	-	1.9	2.5	3.9
All infants and fetuses	155.8	161.7	132.3	144.4	209.3	180.3

Note: Data exclude WA and SA

** Data include country of birth 'not stated'*

Table 2.18 *Selected congenital malformations by maternal country of birth, Australia, 1991-1994 (cont.)*

Congenital malformation	China/ Hong Kong	Philippines	Vietnam	Other Asia	Other countries	All countries*
Total births	11,693	9,577	13,187	23,954	31,813	860,515
			Number			
Anencephalus	3	2	4	3	9	172
Spina bifida	1	3	3	3	9	375
Encephalocele	-	-	-	2	1	73
Hydrocephalus	1	4	8	9	13	329
Transposition of great vessels	2	3	6	6	13	316
Ventricular septal defect	15	16	18	33	70	1,565
Hypoplastic left heart	1	-	2	5	2	176
Cleft palate	4	9	4	10	19	515
Cleft lip	8	12	18	19	32	784
Oesophageal atresia or stenosis	3	2	4	8	8	267
Small intestinal atresia or stenosis	2	3	3	1	5	175
Anorectal atresia or stenosis	6	3	5	11	9	267
Hypospadias	8	10	12	24	69	1,902
Renal agenesis and dysgenesis	2	1	2	9	7	276
Cystic kidney disease	3	1	1	8	10	254
Obstructive defects of renal pelvis and ureter	8	8	4	22	28	753
Congenital dislocation of hip	5	14	10	24	49	1,891
Limb reduction defects	3	3	4	5	15	406
Diaphragmatic hernia	2	1	-	5	8	248
Exomphalos	3	1	5	4	7	138
Gastroschisis	-	-	1	4	1	139
Trisomy 21	17	16	19	18	36	1,153
Trisomy 18	3	5	2	5	15	224
All infants and fetuses	146	155	174	306	552	16,968
			Rate per 10,000 births			
Anencephalus	2.6	2.1	3.0	1.3	2.8	2.0
Spina bifida	0.9	3.1	2.3	1.3	2.8	4.4
Encephalocele	-	-	-	0.8	0.3	0.8
Hydrocephalus	0.9	4.2	6.1	3.8	4.1	3.8
Transposition of great vessels	1.7	3.1	4.5	2.5	4.1	3.7
Ventricular septal defect	12.8	16.7	13.6	13.8	22.0	18.2
Hypoplastic left heart	0.9	-	1.5	2.1	0.6	2.0
Cleft palate	3.4	9.4	3.0	4.2	6.0	6.0
Cleft lip	6.8	12.5	13.6	7.9	10.1	9.1
Oesophageal atresia or stenosis	2.6	2.1	3.0	3.3	2.5	3.1
Small intestinal atresia or stenosis	1.7	3.1	2.3	0.4	1.6	2.0
Anorectal atresia or stenosis	5.1	3.1	3.8	4.6	2.8	3.1
Hypospadias	6.8	10.4	9.1	10.0	21.7	22.1
Renal agenesis and dysgenesis	1.7	1.0	1.5	3.8	2.2	3.2
Cystic kidney disease	2.6	1.0	0.8	3.3	3.1	3.0
Obstructive defects of renal pelvis and ureter	6.8	8.4	3.0	9.2	8.8	8.8
Congenital dislocation of hip	4.3	14.6	7.6	10.0	15.4	22.0
Limb reduction defects	2.6	3.1	3.0	2.1	4.7	4.7
Diaphragmatic hernia	1.7	1.0	-	2.1	2.5	2.9
Exomphalos	2.6	1.0	3.8	1.7	2.2	1.6
Gastroschisis	-	-	0.8	1.7	0.3	1.6
Trisomy 21	14.5	16.7	14.4	7.5	11.3	13.4
Trisomy 18	2.6	5.2	1.5	2.1	4.7	2.6
All infants and fetuses	124.9	161.8	131.9	127.7	173.5	197.2

Note: Data exclude WA and SA

** Data include country of birth 'not stated'*

Table 2.19: ***Live births, stillbirths and total births, States and Territories, 1981-1994***

Year		NSW	Vic	Qld	WA	SA	Tas	ACT	NT	Australia
1981	Live births	47,362	-	38,834	-	19,351	7,188	-	-	112,735
	Stillbirths	353	-	253	-	129	44	-	-	779
	Total births	47,715*	-	39,087	-	19,480	7,232	-	-	113,514
1982	Live births	83,489	59,983	40,540	22,236	19,294	7,002	4,479	-	237,023
	Stillbirths	600	490	225	146	121	48	31	-	1,661
	Total births	84,089	60,473	40,765	22,382	19,415	7,050	4,510	-	238,684
1983	Live births	82,739	60,123	42,000	23,046	19,901	7,028	4,622	-	239,459
	Stillbirths	526	439	262	150	115	49	30	-	1,571
	Total births	83,265	60,562	42,262	23,196	20,016	7,077	4,652	-	241,030
1984	Live births	81,792	59,763	40,356	21,601	20,149	7,098	4,590	-	235,349
	Stillbirths	545	422	245	142	131	45	24	-	1,554
	Total births	82,337	60,185	40,601	21,743	20,280	7,143	4,614	-	236,903
1985	Live births	82,780	61,726	40,275	23,066	19,889	7,213	4,619	-	239,568
	Stillbirths	477	396	251	142	143	52	30	-	1,491
	Total births	83,257	62,122	40,526	23,208	20,032	7,265	4,649	-	241,059
1986	Live births	84,009	60,387	40,166	24,175	19,826	6,911	4,627	3,307	243,408
	Stillbirths	530	403	252	146	125	66	28	35	1,585
	Total births	84,539	60,790	40,418	24,321	19,951	6,977	4,655	3,342	244,993
1987	Live births	85,650	61,642	39,100	23,271	19,345	6,752	4,680	3,519	243,959
	Stillbirths	497	363	231	142	92	29	32	46	1,432
	Total births	86,147	62,005	39,331	23,413	19,437	6,781	4,712	3,565	245,391
1988	Live births	84,268	62,347	40,240	25,123	19,231	6,745	4,817	3,422	246,193
	Stillbirths	523	360	235	120	111	55	36	33	1,473
	Total births	84,791	62,707	40,475	25,243	19,342	6,800	4,853	3,455	247,666
1989	Live births	85,464	64,185	41,714	25,019	19,703	6,788	4,614	3,366	250,853
	Stillbirths	465	411	220	112	132	34	32	45	1,451
	Total births	85,929	64,596	41,934	25,131	19,835	6,822	4,646	3,411	252,304
1990	Live births	90,260	67,158	44,533	25,322	19,981	7,001	4,859	3,534	262,648
	Stillbirths	574	404	245	132	119	45	39	32	1,590
	Total births	90,834	67,562	44,778	25,454	20,100	7,046	4,898	3,566	264,238
1991	Live births	86,220	64,660	44,460	24,815	19,622	6,902	4,490	3,459	254,628
	Stillbirths	692	529	327	192	127	55	41	43	2,006
	Total births	86,912	65,189	44,787	25,007	19,749	6,957	4,531	3,502	256,634
1992	Live births	88,401	65,853	46,307	25,159	20,004	6,975	4,678	3,582	260,959
	Stillbirths	572	447	305	165	148	51	33	46	1,767
	Total births	88,973	66,300	46,612	25,324	20,152	7,026	4,711	3,628	262,726
1993	Live births	87,362	64,323	47,156	25,160	19,844	6,809	4,754	3,505	258,913
	Stillbirths	536	414	292	176	123	47	37	40	1,665
	Total births	87,898	64,737	47,448	25,336	19,967	6,856	4,791	3,545	260,578
1994	Live births	87,488	64,448	47,716	25,237	19,673	6,790	4,747	3,490	259,589
	Stillbirths	496	484	330	188	128	46	37	37	1,746
	Total births	87,984	64,932	48,046	25,425	19,801	6,836	4,784	3,527	261,335
1981-1994	Live births	1,157,284	816,598	593,397	313,230	275,813	97,202	60,576	31,184	3,345,284
	Stillbirths	7,386	5,562	3,673	1,953	1,744	666	430	357	21,771
	Total births	1,164,670	822,160	597,070	315,183	277,557	97,868	61,006	31,541	3,367,055

** Data for NSW were incomplete as some hospitals did not report births in 1981*

Note: Sources of data: 1981-1990 from ABS; 1991-1994 from AIHW National Perinatal Statistics Unit

3.1 Anencephalus

- Anencephalus is a congenital malformation characterised by total or partial absence of the cranial vault, the covering skin and the brain.

- The International Classification of Diseases codes for anencephalus are 740.0-740.2.

- The national rate of anencephalus in births showed a marked decline since 1985, from 5.1 to 1.7 per 10,000 births in 1994, decreasing annually by approximately 0.38 per 10,000 births (Table 3.1, Figure 3.1). During the same period, the number of induced abortions performed before 20 weeks' gestation for anencephalus increased, but notification of these abortions was incomplete.

- In the years 1985-1994, induced abortions were reported in 35.8% of all recorded notifications of anencephalus.

- Among 767 infants with anencephalus and known outcome, 67.1% were stillborn; neonatal deaths were reported in all but 10 liveborn infants. As anencephalus is always a lethal malformation, occasional failure to report the death of a liveborn infant is the most likely explanation for these 10 instances.

- Associated major malformations were reported in 13.1% of the births with anencephalus and 2 infants had a chromosomal abnormality.

- For births in 1990 to 1994, the reported rates for anencephalus were highest in the Northern Territory (5.6 per 10,000 births) and lowest in South Australia (0.8 per 10,000 births) (Table 3.2, Figure 3.2). South Australia's low rate is probably attributable to a statewide screening program for anencephalus and other neural tube defects.

Table 3.1: *Anencephalus by outcome and type of malformation, Australia, 1985-1994*

Outcome	1985	1986	1987	1988	1989	1990	1991	1992	1993	1994	1985-94
						Number					
Live births	47	36	29	26	28	25	15	10	18	18	252
Stillbirths	77	70	63	63	51	47	37	38	43	26	515
Total births*	124	107	94	89	83	73	54	48	64	44	780
Induced abortions	3	29	26	30	39	53	52	62	57	84	435
Neonatal deaths	47	36	29	25	28	25	15	8	15	14	242
						Rate per 10,000 births					
Total births	5.1	4.4	3.8	3.6	3.3	2.8	2.1	1.8	2.5	1.7	3.1
						Number					
Isolated	110	89	85	72	71	68	45	41	56	39	676
Associated	13	18	9	17	12	5	9	7	7	5	102
Chromosomal	1	-	-	-	-	-	-	-	1	-	2
						Rate per 10,000 births					
Isolated	4.6	3.6	3.5	2.9	2.8	2.6	1.8	1.6	2.1	1.5	2.7
Associated	0.5	0.7	0.4	0.7	0.5	0.2	0.4	0.3	0.3	0.2	0.4
Chromosomal	0.0	-	-	-	-	-	-	-	0.0	-	0.0

* *Total includes 'not stated'*

Table 3.2: *Anencephalus, States and Territories, 1990-1994*

Outcome	NSW	Vic	Qld	WA	SA	Tas	ACT	NT	Australia
					Number				
Live births	19	27	23	10	2	2	1	2	86
Stillbirths	52	36	50	35	4	3	3	8	191
Total births*	71	66	73	45	8	6	4	10	283
Induced abortions	54	108	-	54	68	14	8	2	308
					Rate per 10,000 births				
Total births	1.6	2.0	3.2	3.6	0.8	1.7	1.7	5.6	2.2
					Number				
Isolated	61	54	68	41	7	6	4	8	249
Associated	10	12	5	3	1	-	-	2	33
Chromosomal	-	-	-	1	-	-	-	-	1
					Rate per 10,000 births				
Isolated	1.4	1.6	2.9	3.2	0.7	1.7	1.7	4.5	1.9
Associated	0.2	0.4	0.2	0.2	0.1	-	-	1.1	0.3
Chromosomal	-	-	-	0.1	-	-	-	-	0.0

* *Total includes 'not stated'*

Figure 3.1: Anencephalus, Australia, 1985-1994

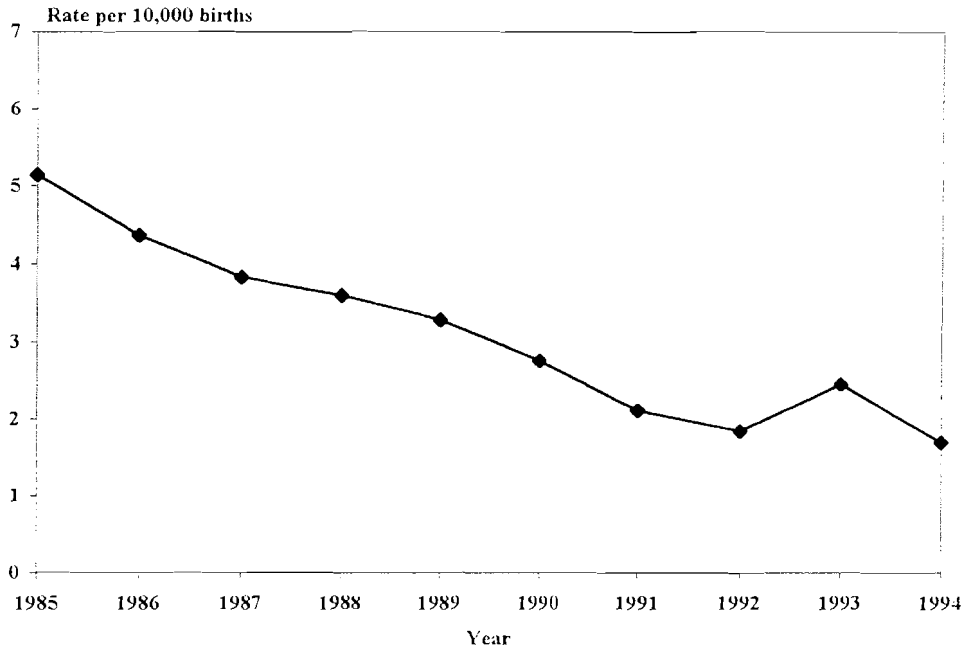
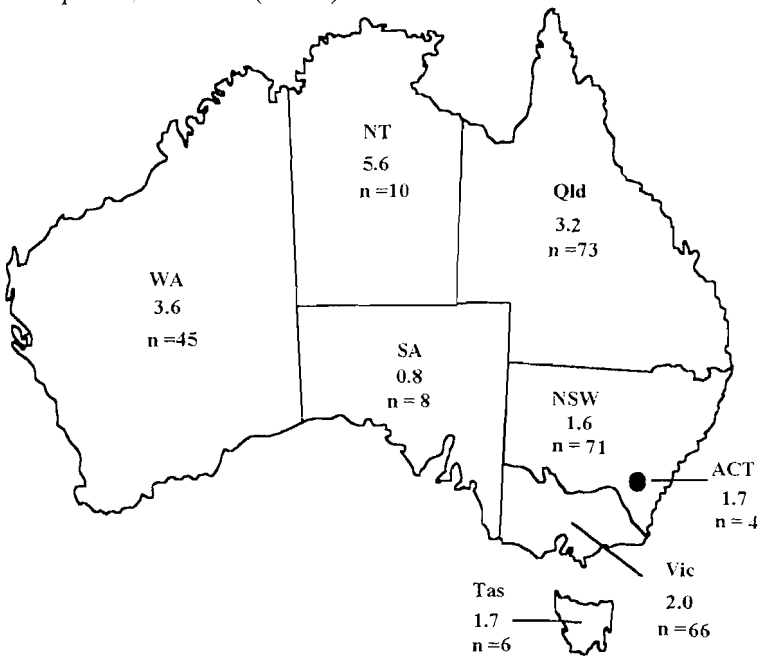


Figure 3.2: Anencephalus, States and Territories, 1990-1994

Australia: 2.2 per 10,000 births (n=283)



3.2 Spina bifida

- Spina bifida is a congenital malformation characterised by herniation or exposure of the spinal cord and/or meninges through an incompletely closed spine. It is not counted as a separate malformation when present with anencephalus; this combination of malformation is often described as craniorachischisis.

- The International Classification of Diseases codes for spina bifida are 741.0-741.9.

- The national rate of spina bifida in births showed a gradual decline from 7.1 per 10,000 births in 1987 to 2.9 per 10,000 births in 1994, decreasing annually by approximately 0.60 per 10,000 births (Table 3.3, Figure 3.3). During the same period, the number of induced abortions performed before 20 weeks' gestation for spina bifida increased, but notification of these abortions was incomplete.

- In the years 1985-1994, induced abortions were reported in 17.2% of all recorded notifications of spina bifida.

- Among 1,420 infants with spina bifida and known outcome, 20.7% were stillborn; 23.2% of liveborn infants died in the neonatal period.

- Associated major malformations were reported in 15.2% of the births with spina bifida and another 3.1% had a chromosomal abnormality.

- For births in 1990 to 1994, the reported rates for spina bifida were highest in Western Australia and Queensland (5.8 and 5.4 per 10,000 births, respectively) and lowest in the Australian Capital Territory and South Australia (3.0 and 3.2 per 10,000 births, respectively) (Table 3.4, Figure 3.4).

Table 3.3: *Spina bifida by outcome and type of malformation, Australia, 1985-1994*

Outcome	1985	1986	1987	1988	1989	1990	1991	1992	1993	1994	1985-94
						Number					
Live births	124	124	136	141	139	129	111	89	72	61	1,126
Stillbirths	30	32	38	29	31	26	32	33	29	14	294
Total births*	154	156	175	171	171	155	145	125	103	77	1,432
Induced abortions	2	13	14	21	27	29	26	45	51	70	298
Neonatal deaths	45	35	32	34	35	29	21	15	7	8	261
						Rate per 10,000 births					
Total births	6.4	6.4	7.1	6.9	6.8	5.9	5.7	4.8	4.0	2.9	5.6
						Number					
Isolated	120	130	151	145	136	122	118	102	80	65	1,169
Associated	28	22	20	22	26	28	23	20	17	12	218
Chromosomal	6	4	4	4	9	5	4	3	6	-	45
						Rate per 10,000 births					
Isolated	5.0	5.3	6.2	5.9	5.4	4.6	4.6	3.9	3.1	2.5	4.6
Associated	1.2	0.9	0.8	0.9	1.0	1.1	0.9	0.8	0.7	0.5	0.9
Chromosomal	0.2	0.2	0.2	0.2	0.4	0.2	0.2	0.1	0.2	-	0.2

** Total includes 'not stated'*

Table 3.4: *Spina bifida, States and Territories, 1990-1994*

Outcome	NSW	Vic	Qld	WA	SA	Tas	ACT	NT	Australia
					Number				
Live births	164	96	105	63	17	7	2	8	462
Stillbirths	31	50	20	11	12	5	5	-	134
Total births*	196	149	126	74	32	13	7	8	605
Induced abortions	41	80	-	25	61	7	5	2	221
					Rate per 10,000 births				
Total births	4.4	4.5	5.4	5.8	3.2	3.7	3.0	4.5	4.6
					Number				
Isolated	146	116	119	60	23	9	7	7	487
Associated	44	27	4	12	9	4	-	-	100
Chromosomal	6	6	3	2	-	-	-	1	18
					Rate per 10,000 births				
Isolated	3.3	3.5	5.1	4.7	2.3	2.6	3.0	3.9	3.7
Associated	1.0	0.8	0.2	0.9	0.9	1.2	-	-	0.8
Chromosomal	0.1	0.2	0.1	0.2	-	-	-	0.6	0.1

** Total includes 'not stated'*

Figure 3.3: Spina bifida, Australia, 1985-1994

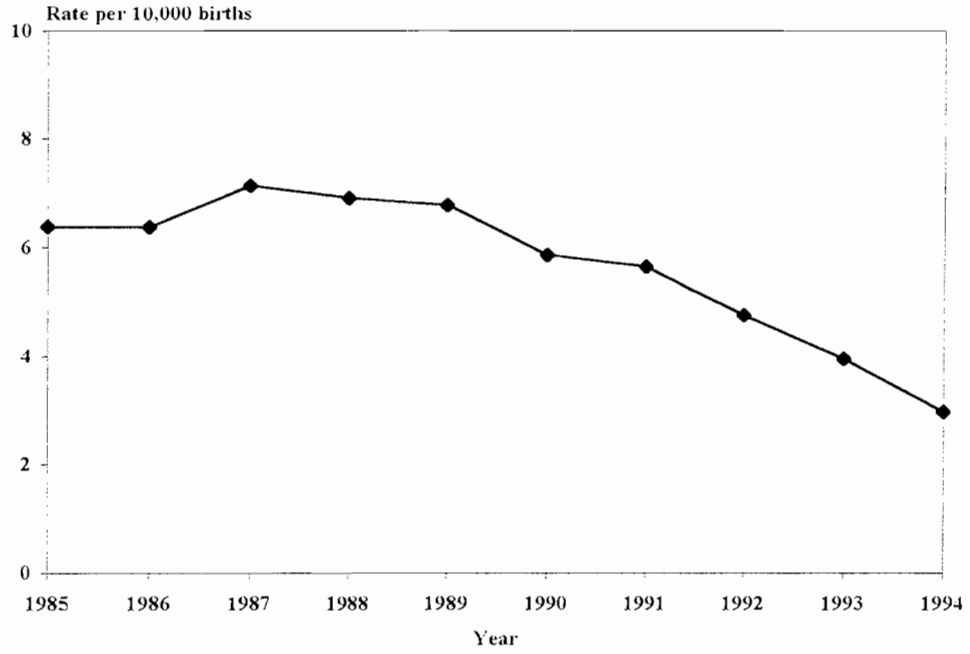
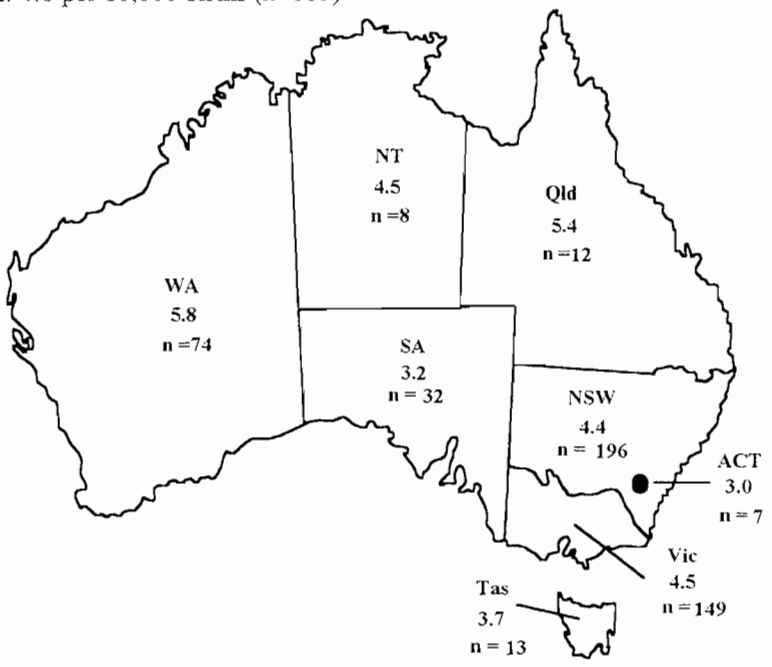


Figure 3.4: Spina bifida, States and Territories, 1990-1994

Australia: 4.6 per 10,000 births (n=605)



3.3 Encephalocele

- Encephalocele is a congenital malformation characterised by herniation of the brain and/or meninges through a defect in the skull. It is not counted as a separate malformation when present with spina bifida. Anencephalus, spina bifida and encephalocele are collectively known as neural tube defects.

- The International Classification of Diseases code for encephalocele is 742.0.

- The national rate of encephalocele in births showed a slow downward trend from a high of 1.7 per 10,000 births in 1986 to a low of 0.7 per 10,000 births in 1993 (Table 3.5, Figure 3.5). During the same period, the number of induced abortions performed before 20 weeks' gestation for encephalocele increased.

- In the years 1985-1994, induced abortions were reported in 17.8% of all recorded notifications of encephalocele.

- Among 257 infants with encephalocele and known outcome, 26.5% were stillborn; 39.7% of liveborn infants died in the neonatal period.

- Associated major malformations were reported in 34.9% of the births with encephalocele and 5 infants had a chromosomal abnormality.

- For births in 1990 to 1994, the reported rates for encephalocele were highest in Victoria and Western Australia (1.3 and 1.1 per 10,000 births, respectively) and lowest in Tasmania and Queensland (0.3 and 0.5 per 10,000 births, respectively) (Table 3.6, Figure 3.6). There was only 1 reported birth each in Tasmania and the Northern Territory.

Table 3.5: *Encephalocele by outcome and type of malformation, Australia, 1985-1994*

Outcome	1985	1986	1987	1988	1989	1990	1991	1992	1993	1994	1985-94
						Number					
Live births	21	35	18	21	17	18	15	19	9	16	189
Stillbirths	8	7	4	9	4	4	8	6	8	10	68
Total births*	29	42	23	30	21	22	23	25	17	26	258
Induced abortions	2	2	3	1	3	7	8	6	9	15	56
Neonatal deaths	9	18	9	10	8	6	4	5	4	2	75
						Rate per 10,000 births					
Total births	1.2	1.7	0.9	1.2	0.8	0.8	0.9	1.0	0.7	1.0	1.0
						Number					
Isolated	21	24	16	23	11	13	15	15	6	19	163
Associated	8	17	7	7	9	9	7	10	10	6	90
Chromosomal	-	1	-	-	1	-	1	-	1	1	5
						Rate per 10,000 births					
Isolated	0.9	1.0	0.7	0.9	0.4	0.5	0.6	0.6	0.2	0.7	0.6
Associated	0.3	0.7	0.3	0.3	0.4	0.3	0.3	0.4	0.4	0.2	0.4
Chromosomal	-	0.0	-	-	0.0	-	0.0	-	0.0	0.0	0.0

** Total includes 'not stated'*

Table 3.6: *Encephalocele, States and Territories, 1990-1994*

Outcome	NSW	Vic	Qld	WA	SA	Tas	ACT	NT	Australia
					Number				
Live births	24	32	8	8	4	1	-	-	77
Stillbirths	11	10	3	6	3	-	2	1	36
Total births*	35	42	11	14	7	1	2	1	113
Induced abortions	7	15	-	9	12	1	1	-	45
					Rate per 10,000 births				
Total births	0.8	1.3	0.5	1.1	0.7	0.3	0.8	0.6	0.9
					Number				
Isolated	23	26	7	7	3	-	2	-	68
Associated	11	15	3	7	4	1	-	1	42
Chromosomal	1	1	1	-	-	-	-	-	3
					Rate per 10,000 births				
Isolated	0.5	0.8	0.3	0.6	0.3	-	0.8	-	0.5
Associated	0.2	0.5	0.1	0.6	0.4	0.3	-	0.6	0.3
Chromosomal	0.0	0.0	0.0	-	-	-	-	-	0.0

** Total includes 'not stated'*

Figure 3.5: Encephalocele, Australia, 1985-1994

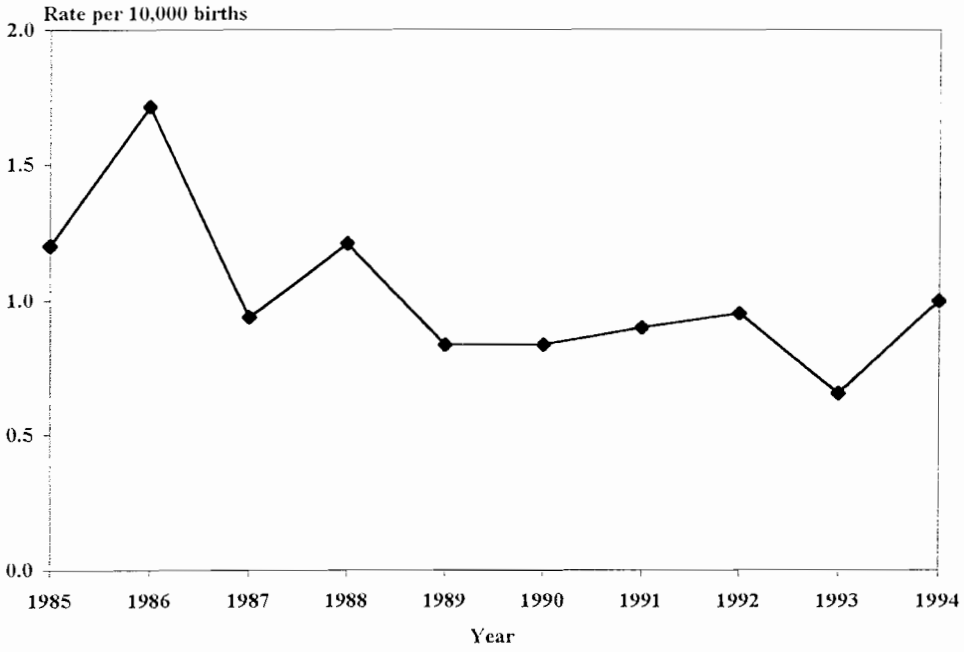
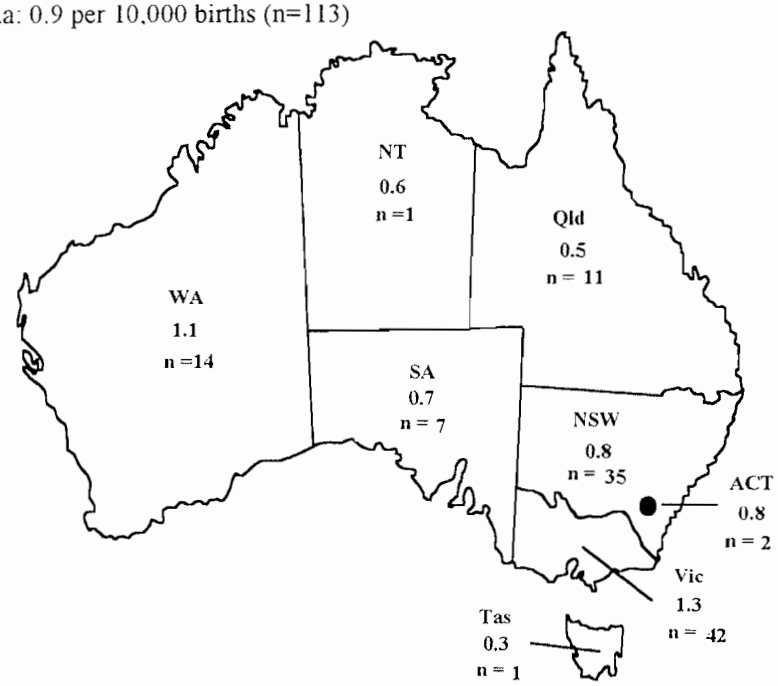


Figure 3.6: Encephalocele, States and Territories, 1990-1994

Australia: 0.9 per 10,000 births (n=113)



3.4 Microcephalus

- Microcephalus is a congenital malformation characterised by a reduced brain size and head circumference. The head circumference is more than three standard deviations below the mean measurement of infants of the same gestational age.

- The International Classification of Diseases code for microcephalus is 742.1.

- The national rate of microcephalus in births was quite varied, showing a downward trend since 1989 from 2.3 per 10,000 births to 1.1 per 10,000 births in 1994 (Table 3.7, Figure 3.7).

- In the years 1985-1994, induced abortions were reported in 1.6% of all recorded notifications of microcephalus.

- Among 415 infants with microcephalus and known outcome, 8.0% were stillborn; 28.3% of liveborn infants died in the neonatal period.

- Associated major malformations were reported in 34.2% of the births with microcephalus and another 22.0% had a chromosomal abnormality.

- For births in 1990 to 1994, the reported rates for microcephalus were highest in the Northern Territory (3.4 per 10,000 births) and lowest in Tasmania and the Australian Capital Territory (0.6 and 0.8 per 10,000 births, respectively) (Table 3.8, Figure 3.8).

Table 3.7: *Microcephalus by outcome and type of malformation, Australia, 1985-1994*

Outcome	1985	1986	1987	1988	1989	1990	1991	1992	1993	1994	1985-94
						Number					
Live births	47	42	30	27	55	45	39	32	38	27	382
Stillbirths	5	4	3	6	2	4	4	2	1	2	33
Total births*	52	46	33	33	57	49	43	34	42	29	418
Induced abortions	1	-	-	-	1	-	2	-	-	3	7
Neonatal deaths	21	17	8	8	14	11	9	7	7	6	108
						Rate per 10,000 births					
Total births	2.2	1.9	1.3	1.3	2.3	1.9	1.7	1.3	1.6	1.1	1.6
						Number					
Isolated	23	16	13	14	27	27	16	19	15	13	183
Associated	15	17	10	10	18	10	18	11	21	13	143
Chromosomal	14	13	10	9	12	12	9	4	6	3	92
						Rate per 10,000 births					
Isolated	1.0	0.7	0.5	0.6	1.1	1.0	0.6	0.7	0.6	0.5	0.7
Associated	0.6	0.7	0.4	0.4	0.7	0.4	0.7	0.4	0.8	0.5	0.6
Chromosomal	0.6	0.5	0.4	0.4	0.5	0.5	0.4	0.2	0.2	0.1	0.4

* *Total includes 'not stated'*

Table 3.8: *Microcephalus, States and Territories, 1990-1994*

Outcome	NSW	Vic	Qld	WA	SA	Tas	ACT	NT	Australia
					Number				
Live births	64	56	28	16	8	2	2	5	181
Stillbirths	3	2	1	4	2	-	-	1	13
Total births*	70	58	29	20	10	2	2	6	197
Induced abortions	-	-	-	4	1	-	-	-	5
					Rate per 10,000 births				
Total births	1.6	1.8	1.3	1.6	1.0	0.6	0.8	3.4	1.5
					Number				
Isolated	29	36	10	6	4	2	2	1	90
Associated	29	16	6	13	4	-	-	5	73
Chromosomal	12	6	13	1	2	-	-	-	34
					Rate per 10,000 births				
Isolated	0.7	1.1	0.4	0.5	0.4	0.6	0.8	0.6	0.7
Associated	0.7	0.5	0.3	1.0	0.4	-	-	2.8	0.6
Chromosomal	0.3	0.2	0.6	0.1	0.2	-	-	-	0.3

* *Total includes 'not stated'*

Figure 3.7: Microcephalus, Australia, 1985-1994

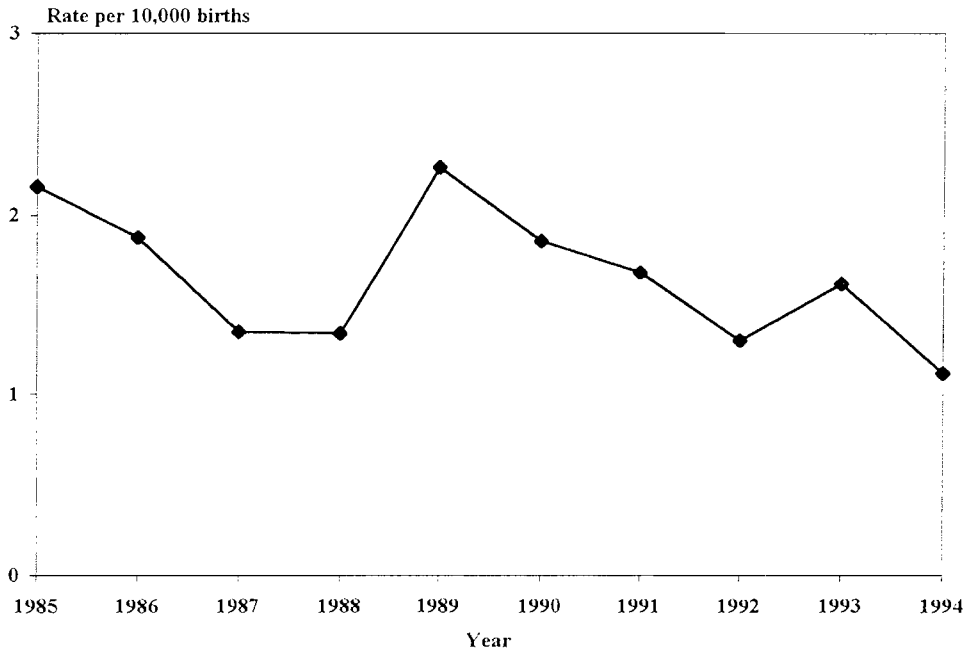
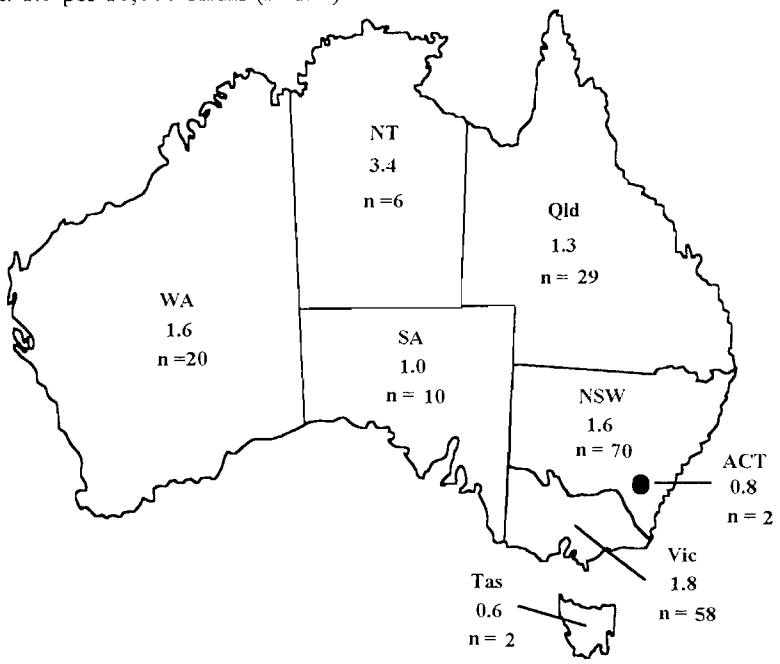


Figure 3.8: Microcephalus, States and Territories, 1990-1994

Australia: 1.5 per 10,000 births (n=197)



3.5 Hydrocephalus

- Hydrocephalus is a congenital malformation characterised by dilation of the ventricular system, not associated with primary brain atrophy, with or without enlargement of the head, and diagnosed before birth or during the first week of life. It is not counted as a separate malformation when present with encephalocele or open spina bifida.

- The International Classification of Diseases code for hydrocephalus is 742.3.

- There was no clear trend in the national rate of hydrocephalus in births during 1985 to 1994, with an overall rate of 3.8 per 10,000 births (Table 3.9, Figure 3.9). Relatively more induced abortions performed before 20 weeks' gestation for hydrocephalus were reported in 1991 to 1994 than in earlier years.

- In the years 1985-1994, induced abortions were reported in 12.1% of all recorded notifications of hydrocephalus.

- Among 951 infants with hydrocephalus and known outcome, 36.1% were stillborn; 29.3% of liveborn infants died in the neonatal period.

- Associated major malformations were reported in 29.2% of the births with hydrocephalus and another 7.8% had a chromosomal abnormality.

- For births in 1990 to 1994, the reported rates for hydrocephalus were highest in the Australian Capital Territory and the Northern Territory (5.9 and 5.6 per 10,000 births, respectively) and lowest in South Australia (3.0 per 10,000 births) (Table 3.10, Figure 3.10).

Table 3.9: *Hydrocephalus by outcome and type of malformation, Australia, 1985-1994*

Outcome	1985	1986	1987	1988	1989	1990	1991	1992	1993	1994	1985-94
						Number					
Live births	62	46	55	63	63	82	56	54	65	62	608
Stillbirths	41	26	34	33	30	40	33	40	33	33	343
Total births*	103	72	89	97	94	123	89	97	100	96	960
Induced abortions	-	8	6	7	8	8	17	17	30	31	132
Neonatal deaths	24	19	24	30	12	15	14	15	15	10	178
						Rate per 10,000 births					
Total births	4.3	2.9	3.6	3.9	3.7	4.7	3.5	3.7	3.8	3.7	3.8
						Number					
Isolated	62	42	63	56	58	88	55	55	63	63	605
Associated	33	23	22	32	32	26	31	31	26	24	280
Chromosomal	8	7	4	9	4	9	3	11	11	9	75
						Rate per 10,000 births					
Isolated	2.6	1.7	2.6	2.3	2.3	3.3	2.1	2.1	2.4	2.4	2.4
Associated	1.4	0.9	0.9	1.3	1.3	1.0	1.2	1.2	1.0	0.9	1.1
Chromosomal	0.3	0.3	0.2	0.4	0.2	0.3	0.1	0.4	0.4	0.3	0.3

** Total includes 'not stated'*

Table 3.10: *Hydrocephalus, States and Territories, 1990-1994*

Outcome	NSW	Vic	Qld	WA	SA	Tas	ACT	NT	Australia
					Number				
Live births	124	86	50	25	15	9	4	6	319
Stillbirths	36	70	24	17	13	6	10	3	179
Total births*	162	157	75	42	30	15	14	10	505
Induced abortions	18	51	1	14	15	2	2	-	103
					Rate per 10,000 births				
Total births	3.7	4.8	3.2	3.3	3.0	4.3	5.9	5.6	3.9
					Number				
Isolated	105	94	49	26	19	12	10	9	324
Associated	49	43	18	16	8	1	2	1	138
Chromosomal	8	20	8	-	3	2	2	-	43
					Rate per 10,000 births				
Isolated	2.4	2.9	2.1	2.1	1.9	3.5	4.2	5.1	2.5
Associated	1.1	1.3	0.8	1.3	0.8	0.3	0.8	0.6	1.1
Chromosomal	0.2	0.6	0.3	-	0.3	0.6	0.8	-	0.3

** Total includes 'not stated'*

Figure 3.9: Hydrocephalus, Australia, 1985-1994

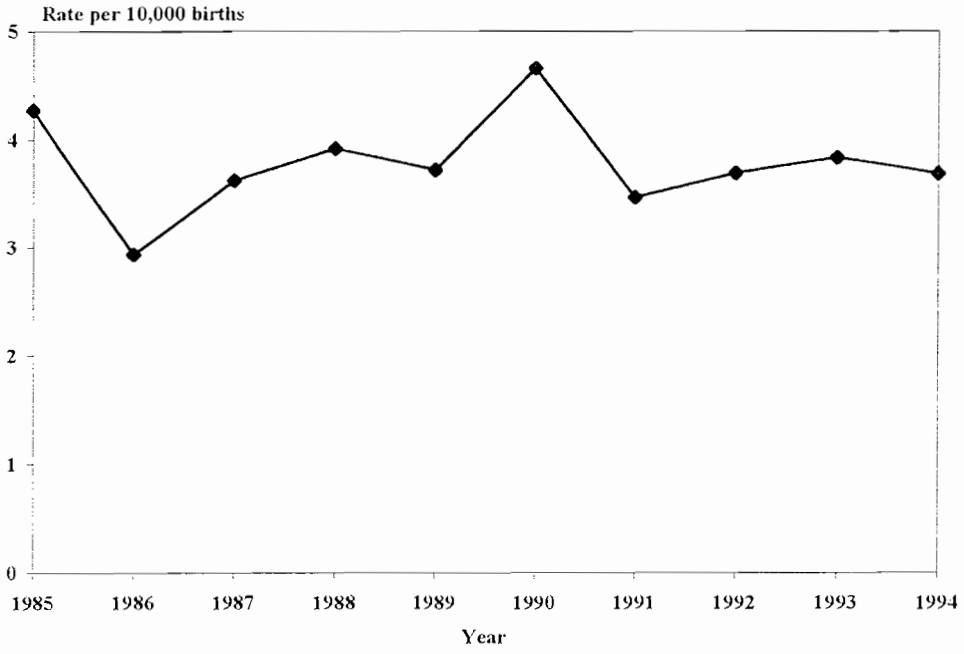
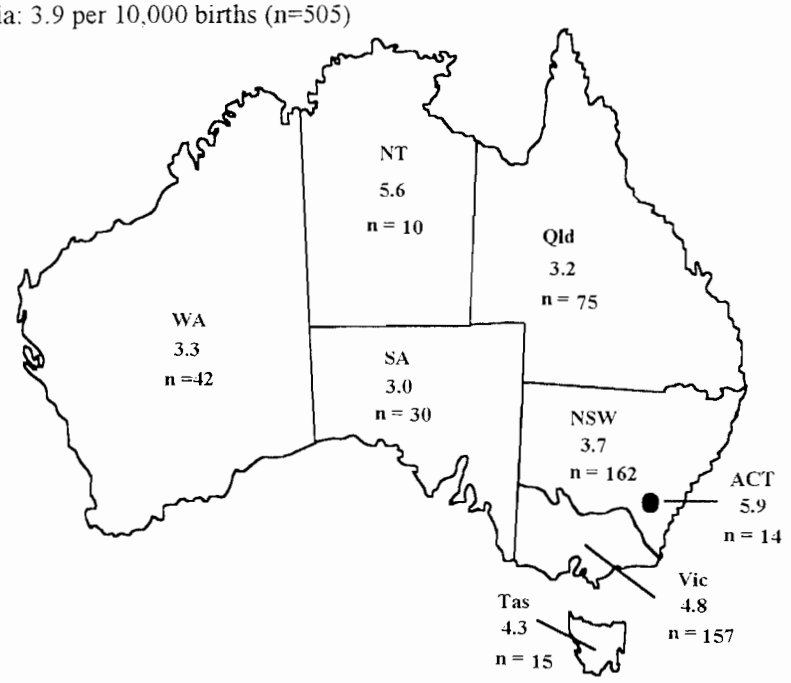


Figure 3.10: Hydrocephalus, States and Territories, 1990-1994

Australia: 3.9 per 10,000 births (n=505)



3.6 Transposition of great vessels

- Transposition of the great vessels is a congenital heart defect usually characterised by the aorta arising from the right ventricle and the pulmonary artery from the left ventricle.
- The International Classification of Diseases code for transposition of the great vessels is 745.1.
- The national rate of transposition of the great vessels showed no clear trend between 1985 and 1994 and was 3.8 per 10,000 births for this period (Table 3.11, Figure 3.11).
- In the years 1985-1994, induced abortions were reported in 0.7% of all recorded notifications of transposition of the great vessels.
- Among 956 infants with transposition of the great vessels and known outcome, 4.1% were stillborn; 19.8% of liveborn infants died in the neonatal period.
- Associated major malformations were reported in 28.5% of the births with transposition of the great vessels and another 4.6% had a chromosomal abnormality.
- For births in 1990 to 1994, the reported rates for transposition of the great vessels were highest in South Australia and Victoria (4.8 and 4.7 per 10,000 births, respectively) and lowest in the Australian Capital Territory (2.1 per 10,000 births) (Table 3.12, Figure 3.12).

Table 3.11: *Transposition of great vessels by outcome and type of malformation, Australia, 1985-1994*

Outcome	1985	1986	1987	1988	1989	1990	1991	1992	1993	1994	1985-94
						Number					
Live births	85	80	92	114	98	83	82	100	87	96	917
Stillbirths	6	1	4	1	4	6	6	6	2	3	39
Total births*	91	81	96	115	102	89	88	106	89	99	956
Induced abortions	-	-	-	2	-	-	1	1	1	2	7
Neonatal deaths	24	21	20	31	23	12	19	12	14	6	182
						Rate per 10,000 births					
Total births	3.8	3.3	3.9	4.6	4.0	3.4	3.4	4.0	3.4	3.8	3.8
						Number					
Isolated	57	53	67	73	65	58	49	84	60	74	640
Associated	28	23	25	37	34	25	33	19	24	24	272
Chromosomal	6	5	4	5	3	6	6	3	5	1	44
						Rate per 10,000 births					
Isolated	2.4	2.2	2.7	2.9	2.6	2.2	1.9	3.2	2.3	2.8	2.5
Associated	1.2	0.9	1.0	1.5	1.3	0.9	1.3	0.7	0.9	0.9	1.1
Chromosomal	0.2	0.2	0.2	0.2	0.1	0.2	0.2	0.1	0.2	0.0	0.2

* *Total includes 'not stated'*

Table 3.12: *Transposition of great vessels, States and Territories, 1990-1994*

Outcome	NSW	Vic	Qld	WA	SA	Tas	ACT	NT	Australia
					Number				
Live births	130	144	75	38	47	9	5	5	453
Stillbirths	4	10	4	1	1	3	-	-	23
Total births*	134	154	79	39	48	12	5	5	476
Induced abortions	1	3	-	1	-	-	-	-	5
					Rate per 10,000 births				
Total births	3.0	4.7	3.4	3.1	4.8	3.5	2.1	2.8	3.6
					Number				
Isolated	80	110	66	25	33	8	3	3	328
Associated	45	39	12	12	13	3	1	1	126
Chromosomal	9	5	1	2	2	1	1	1	22
					Rate per 10,000 births				
Isolated	1.8	3.3	2.9	2.0	3.3	2.3	1.3	1.7	2.5
Associated	1.0	1.2	0.5	0.9	1.3	0.9	0.4	0.6	1.0
Chromosomal	0.2	0.2	0.0	0.2	0.2	0.3	0.4	0.6	0.2

* *Total includes 'not stated'*

Figure 3.11: Transposition of great vessels, Australia, 1985-1994

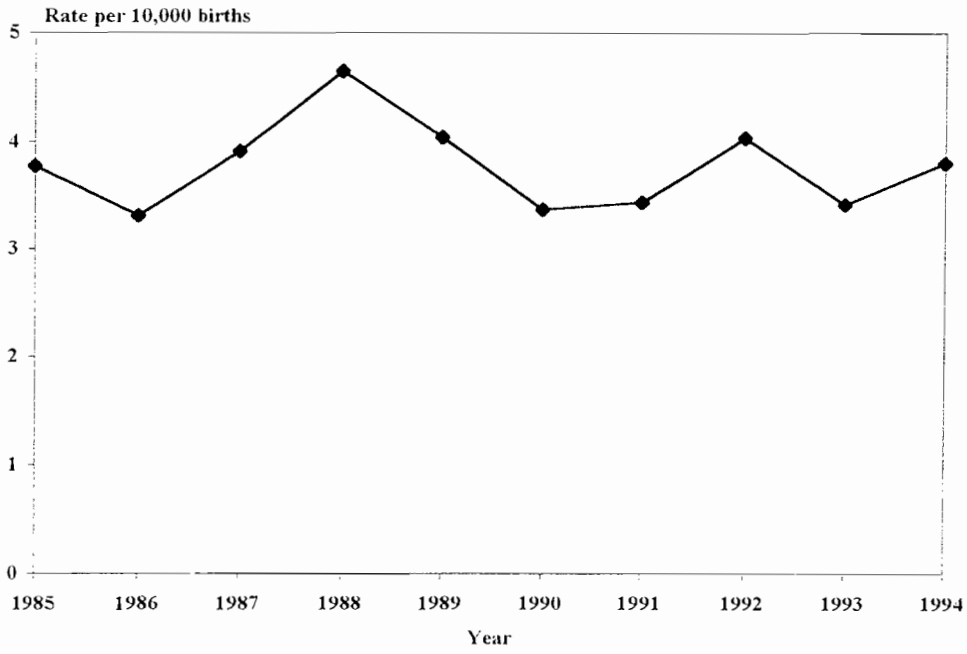
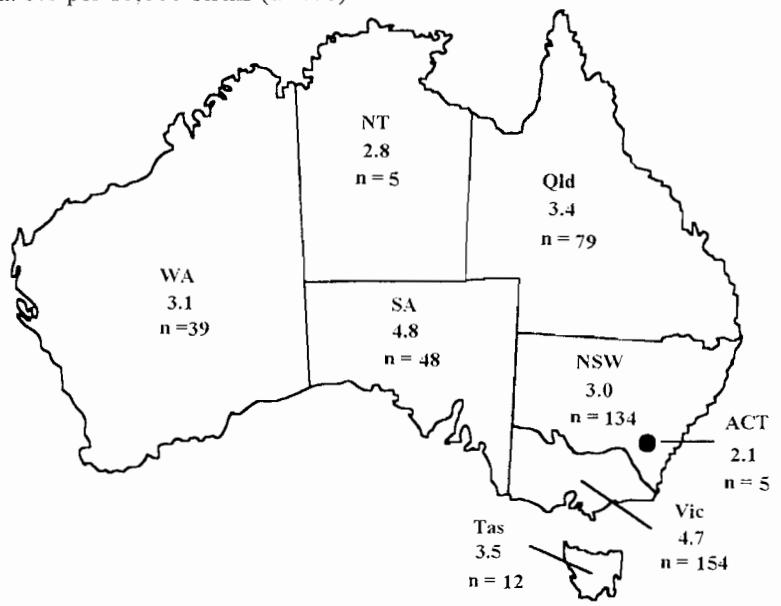


Figure 3.12: Transposition of great vessels, States and Territories, 1990-1994

Australia: 3.6 per 10,000 births (n=476)



3.7 Ventricular septal defect

- Ventricular septal defect is an abnormal communication between the ventricles of the heart, usually characterised by a heart murmur and sometimes by spontaneous closure in early childhood.

- The International Classification of Diseases code for ventricular septal defect is 745.4.

- The national rate of ventricular septal defect in births showed a very gradual increase from a low of 12.8 per 10,000 births in 1986 to a high of 19.1 per 10,000 births in 1993, increasing annually by approximately 0.9 per 10,000 births (Table 3.13, Figure 3.13).

- In the years 1985-1994, induced abortions were reported in 1.7% of all recorded notifications of ventricular septal defect.

- Among 4,119 infants with ventricular septal defect and known outcome, 6.0% were stillborn; 10.7% of liveborn infants died in the neonatal period.

- Associated major malformations were reported in 34.3% of the births with ventricular septal defect and another 12.6% had a chromosomal abnormality.

- For births in 1990 to 1994, the reported rates for ventricular septal defect were highest in Victoria, the Northern Territory and South Australia (23.2, 23.1 and 22.5 per 10,000 births, respectively) and lowest in the Australian Capital Territory and Tasmania (6.7 and 7.5 per 10,000 births, respectively) (Table 3.14, Figure 3.14).

Table 3.13: *Ventricular septal defect by outcome and type of malformation, Australia, 1985-1994*

Outcome	1985	1986	1987	1988	1989	1990	1991	1992	1993	1994	1985-94
						Number					
Live births	288	300	340	339	370	465	389	463	465	451	3,870
Stillbirths	23	14	24	23	24	27	24	34	27	29	249
Total births*	311	314	364	362	394	498	416	499	497	481	4,136
Induced abortions	1	1	2	5	7	6	9	7	13	19	70
Neonatal deaths	73	39	51	48	43	39	37	28	29	27	414
						Rate per 10,000 births					
Total births	12.9	12.8	14.8	14.6	15.6	18.8	16.2	19.0	19.1	18.5	16.3
						Number					
Isolated	144	172	185	167	200	263	228	290	279	268	2,196
Associated	121	106	138	150	137	166	140	161	156	143	1,418
Chromosomal	46	36	41	45	57	69	48	48	62	70	522
						Rate per 10,000 births					
Isolated	6.0	7.0	7.5	6.7	7.9	10.0	8.9	11.0	10.7	10.3	8.7
Associated	5.0	4.3	5.6	6.1	5.4	6.3	5.5	6.1	6.0	5.5	5.6
Chromosomal	1.9	1.5	1.7	1.8	2.3	2.6	1.9	1.8	2.4	2.7	2.1

** Total includes 'not stated'*

Table 3.14: *Ventricular septal defect, States and Territories, 1990-1994*

Outcome	NSW	Vic	Qld	WA	SA	Tas	ACT	NT	Australia
					Number				
Live births	665	703	402	186	201	21	14	41	2,233
Stillbirths	28	54	18	13	21	5	2	-	141
Total births*	701	761	422	200	224	26	16	41	2,391
Induced abortions	-	31	-	8	13	1	1	-	54
					Rate per 10,000 births				
Total births	15.9	23.2	18.2	15.8	22.5	7.5	6.7	23.1	18.3
					Number				
Isolated	355	424	302	81	117	18	8	23	1,328
Associated	252	243	88	81	79	5	5	13	766
Chromosomal	94	94	32	38	28	3	3	5	297
					Rate per 10,000 births				
Isolated	8.0	12.9	13.1	6.4	11.7	5.2	3.4	12.9	10.2
Associated	5.7	7.4	3.8	6.4	7.9	1.4	2.1	7.3	5.9
Chromosomal	2.1	2.9	1.4	3.0	2.8	0.9	1.3	2.8	2.3

** Total includes 'not stated'*

Figure 3.13: Ventricular septal defect, Australia, 1985-1994

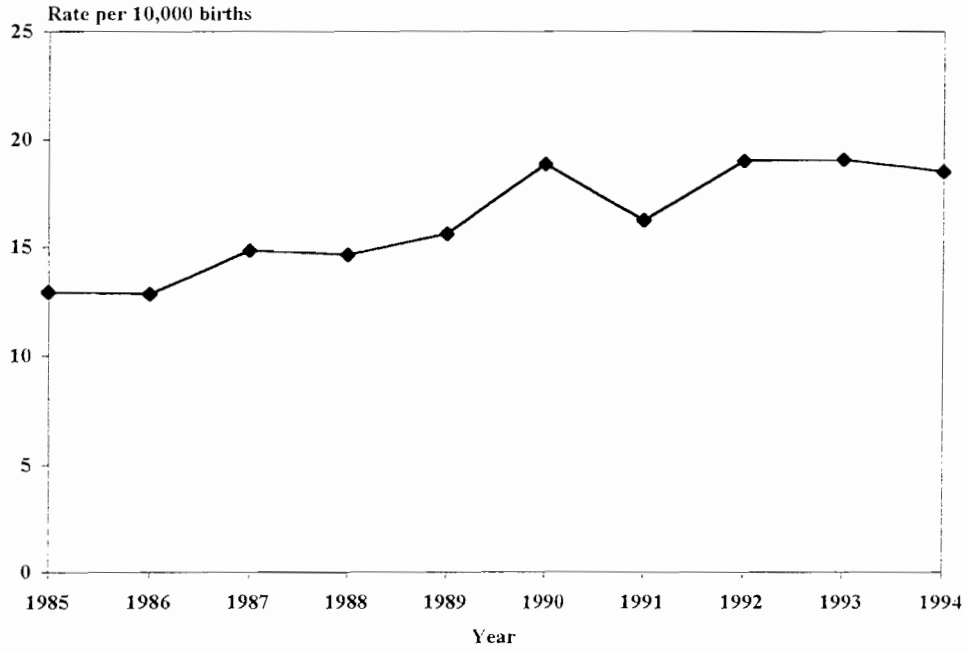
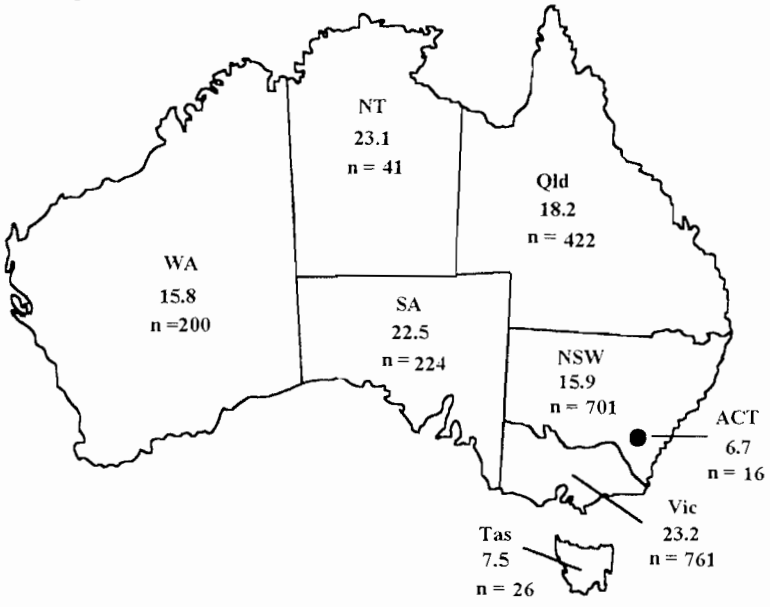


Figure 3.14: Ventricular septal defect, States and Territories, 1990-1994

Australia: 18.3 per 10,000 births (n=2,391)



3.8 Hypoplastic left heart

- Hypoplastic left heart is a congenital malformation resulting from an obstructive valvular and vascular lesion of the left side of the heart with varying degrees of hypoplasia of the left ventricle.
- The International Classification of Diseases code for hypoplastic left heart is 746.7.
- The national rate of hypoplastic left heart in births showed a decline from 2.8 per 10,000 births in 1991 to 1.6 per 10,000 births in 1994 (Table 3.15, Figure 3.15).
- In the years 1985-1994, induced abortions were reported in 3.7% of all recorded notifications of hypoplastic left heart.
- Among 549 infants with hypoplastic left heart and known outcome, 7.8% were stillborn; 73.5% of liveborn infants died in the neonatal period. As this is a lethal condition, there may be incomplete reporting of neonatal deaths, or the initial diagnosis reported on perinatal forms may sometimes be incorrect.
- Associated major malformations were reported in 14.9% of the births with hypoplastic left heart and another 6.4% had a chromosomal abnormality.
- For births in 1990 to 1994, the reported rates for hypoplastic left heart were highest in the Northern Territory, South Australia and Western Australia (3.4, 3.0 and 2.9 per 10,000 births, respectively) and lowest in the Australian Capital Territory (0.8 per 10,000 births) (Table 3.16, Figure 3.16).

Table 3.15: *Hypoplastic left heart by outcome and type of malformation, Australia, 1985-1994*

Outcome	1985	1986	1987	1988	1989	1990	1991	1992	1993	1994	1985-94
						Number					
Live births	47	54	50	56	48	52	67	46	53	33	506
Stillbirths	-	1	2	2	3	5	4	9	7	10	43
Total births*	47	55	52	58	51	57	72	55	60	43	550
Induced abortions	-	-	-	-	1	3	6	2	3	6	21
Neonatal deaths	41	48	44	45	39	43	37	25	32	18	372
						Rate per 10,000 births					
Total births	1.9	2.2	2.1	2.3	2.0	2.2	2.8	2.1	2.3	1.6	2.2
						Number					
Isolated	38	51	43	49	39	43	53	42	40	35	433
Associated	4	2	8	8	6	10	12	8	18	6	82
Chromosomal	5	2	1	1	6	4	7	5	2	2	35
						Rate per 10,000 births					
Isolated	1.6	2.1	1.8	2.0	1.5	1.6	2.1	1.6	1.5	1.3	1.7
Associated	0.2	0.1	0.3	0.3	0.2	0.4	0.5	0.3	0.7	0.2	0.3
Chromosomal	0.2	0.1	0.0	0.0	0.2	0.2	0.3	0.2	0.1	0.1	0.1

** Total includes 'not stated'*

Table 3.16: *Hypoplastic left heart, States and Territories, 1990-1994*

Outcome	NSW	Vic	Qld	WA	SA	Tas	ACT	NT	Australia
					Number				
Live births	72	65	45	30	28	4	1	6	251
Stillbirths	5	15	5	6	2	1	1	-	35
Total births*	77	80	50	37	30	5	2	6	287
Induced abortions	-	10	-	5	4	-	1	-	20
					Rate per 10,000 births				
Total births	1.7	2.4	2.2	2.9	3.0	1.4	0.8	3.4	2.2
					Number				
Isolated	57	58	43	23	21	3	2	6	213
Associated	18	13	6	8	8	1	-	-	54
Chromosomal	2	9	1	6	1	1	-	-	20
					Rate per 10,000 births				
Isolated	1.3	1.8	1.9	1.8	2.1	0.9	0.8	3.4	1.6
Associated	0.4	0.4	0.3	0.6	0.8	0.3	-	-	0.4
Chromosomal	0.0	0.3	0.0	0.5	0.1	0.3	-	-	0.2

** Total includes 'not stated'*

Figure 3.15: Hypoplastic left heart, Australia, 1985-1994

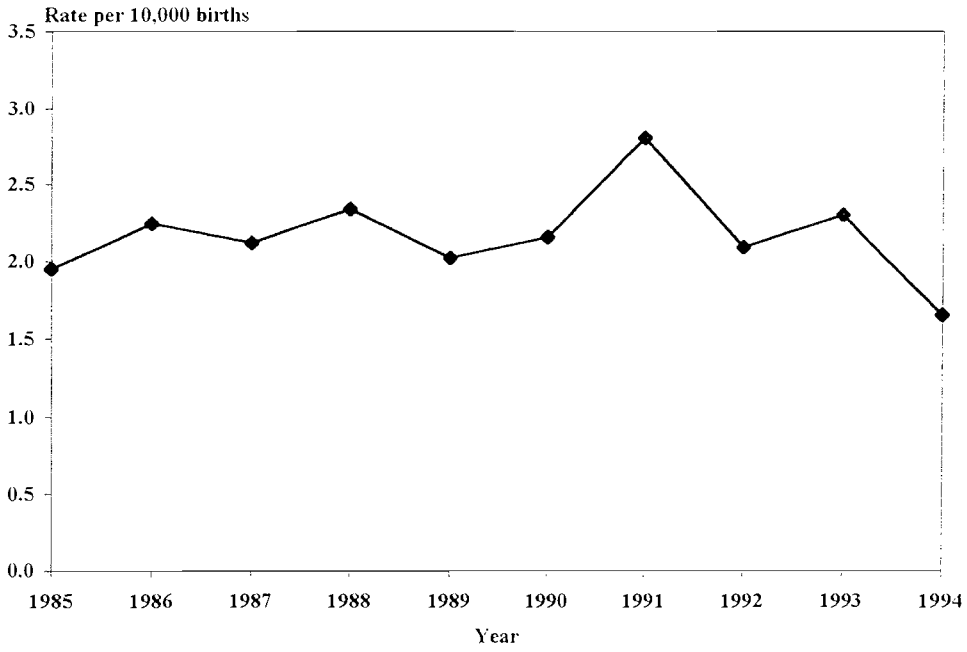
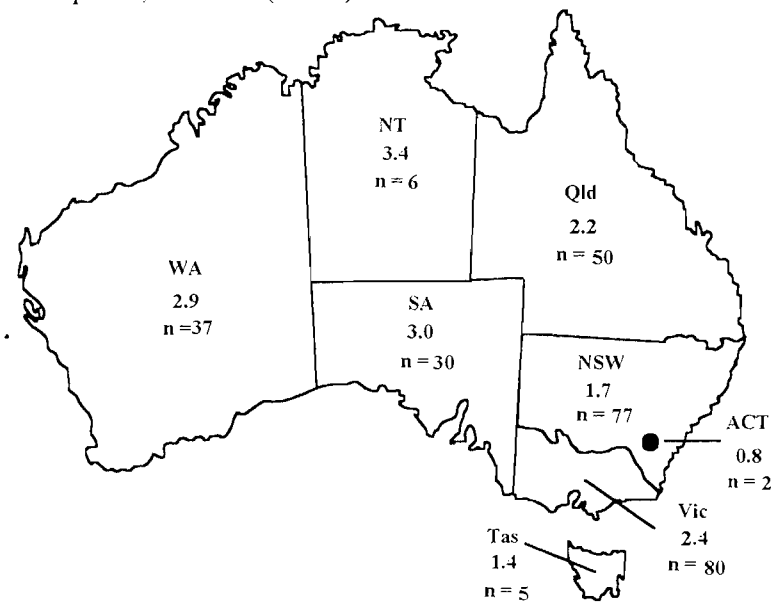


Figure 3.16: Hypoplastic left heart, States and Territories, 1990-1994

Australia: 2.2 per 10,000 births (n=287)



3.9 Coarctation of aorta

- Coarctation of the aorta is a congenital malformation resulting from narrowing of the aorta, either proximal or distal to the ductus arteriosus.
- The International Classification of Diseases code for coarctation of the aorta is 747.1.
- The national rate of coarctation of the aorta in births showed a decline since the late 1980s from 3.8 per 10,000 births in 1988 to 2.3 per 10,000 births in 1994 (Table 3.17, Figure 3.17).
- In the years 1985-1994, induced abortions were reported in 1.2% of all recorded notifications of coarctation of the aorta.
- Among 767 infants with coarctation of the aorta and known outcome, 3.9% were stillborn; 18.9% of liveborn infants died in the neonatal period.
- Associated major malformations were reported in 37.9% of the births with coarctation of the aorta and another 8.1% had a chromosomal abnormality.
- For births in 1990 to 1994, the reported rates for coarctation of the aorta were highest in Victoria (4.6 per 10,000 births) and lowest in the Australian Capital Territory, Tasmania and the Northern Territory (0.4, 0.9 and 1.1 per 10,000 births, respectively) (Table 3.18, Figure 3.18). There was only 1 reported birth in the Australian Capital Territory.

Table 3.17: *Coarctation of aorta by outcome and type of malformation, Australia, 1985-1994*

Outcome	1985	1986	1987	1988	1989	1990	1991	1992	1993	1994	1985-94
						Number					
Live births	54	79	87	92	78	91	57	76	66	57	737
Stillbirths	3	1	2	1	3	2	4	4	6	4	30
Total births*	57	80	89	93	81	94	62	80	73	61	770
Induced abortions	-	1	-	-	1	-	1	2	-	4	9
Neonatal deaths	21	21	20	21	11	14	7	7	10	7	139
						Rate per 10,000 births					
Total births	2.4	3.3	3.6	3.8	3.2	3.6	2.4	3.0	2.8	2.3	3.0
						Number					
Isolated	28	47	53	49	46	51	32	45	34	31	416
Associated	23	26	31	41	29	34	27	31	29	21	292
Chromosomal	6	7	5	3	6	9	3	4	10	9	62
						Rate per 10,000 births					
Isolated	1.2	1.9	2.2	2.0	1.8	1.9	1.2	1.7	1.3	1.2	1.6
Associated	1.0	1.1	1.3	1.7	1.1	1.3	1.1	1.2	1.1	0.8	1.2
Chromosomal	0.2	0.3	0.2	0.1	0.2	0.3	0.1	0.2	0.4	0.3	0.2

** Total includes 'not stated'*

Table 3.18: *Coarctation of aorta, States and Territories, 1990-1994*

Outcome	NSW	Vic	Qld	WA	SA	Tas	ACT	NT	Australia
					Number				
Live births	98	141	48	34	21	2	1	2	347
Stillbirths	5	8	2	1	3	1	-	-	20
Total births*	105	150	50	35	24	3	1	2	370
Induced abortions	-	2	-	2	3	-	-	-	7
					Rate per 10,000 births				
Total births	2.4	4.6	2.2	2.8	2.4	0.9	0.4	1.1	2.8
					Number				
Isolated	52	78	37	13	10	2	-	1	193
Associated	41	58	10	18	12	1	1	1	142
Chromosomal	12	14	3	4	2	-	-	-	35
					Rate per 10,000 births				
Isolated	1.2	2.4	1.6	1.0	1.0	0.6	-	0.6	1.5
Associated	0.9	1.8	0.4	1.4	1.2	0.3	0.4	0.6	1.1
Chromosomal	0.3	0.4	0.1	0.3	0.2	-	-	-	0.3

** Total includes 'not stated'*

Figure 3.17: Coarctation of aorta, Australia, 1985-1994

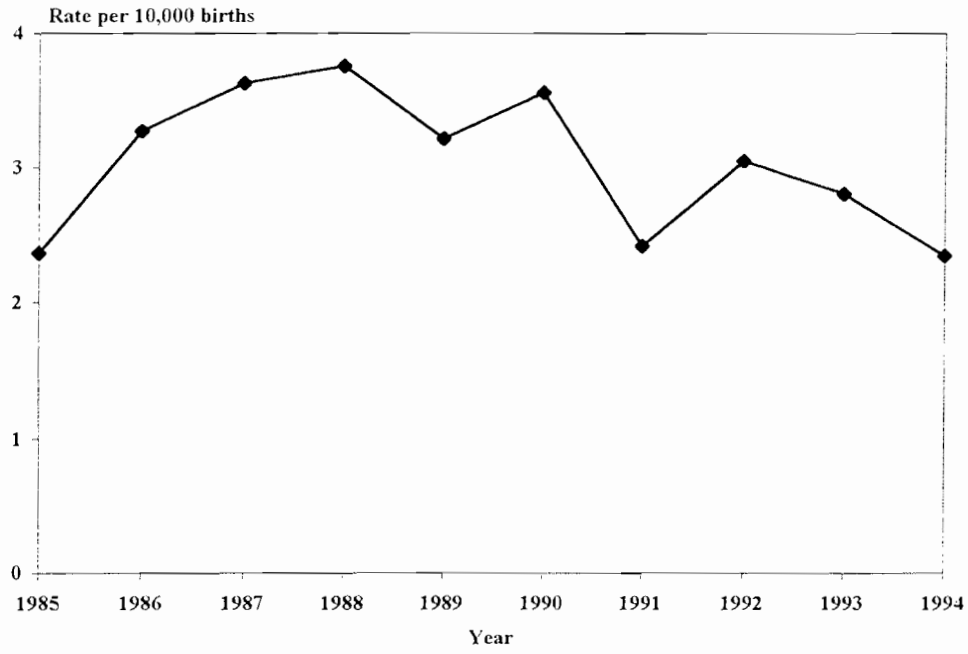
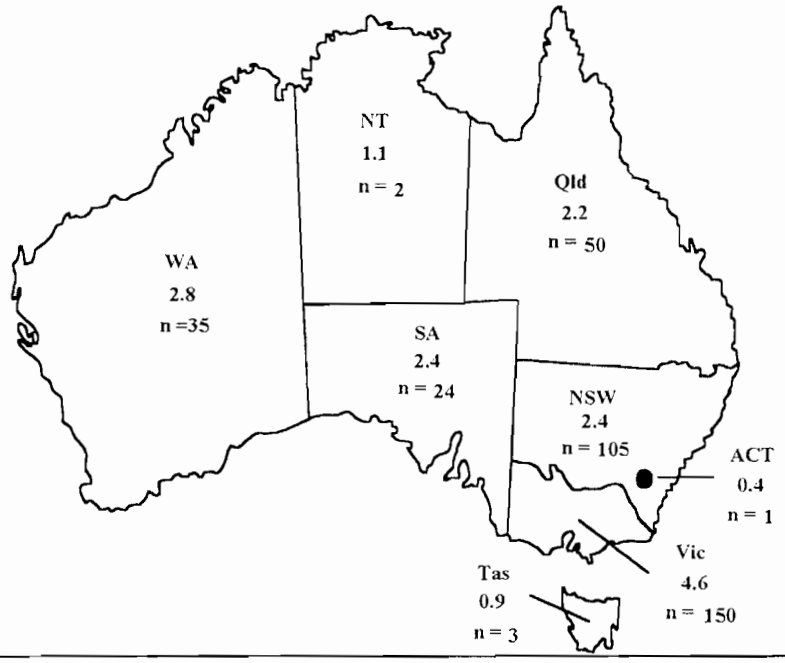


Figure 3.18: Coarctation of aorta, States and Territories, 1990-1994

Australia: 2.8 per 10,000 births (n=370)



3.10 Cleft palate

- Cleft palate is a congenital malformation characterised by a fissure defect of the hard and/or soft palate behind the foramen incisivum without cleft lip.

- The International Classification of Diseases code for cleft palate is 749.0.

- The national rate of cleft palate in births showed very little change between 1985 and 1994, with an overall rate of 5.7 per 10,000 births in this period (Table 3.19, Figure 3.19). During the 1990's 25 of 27 induced abortions performed before 20 weeks' gestation for cleft palate occurred.

- In the years 1985-1994, induced abortions were reported in 1.8% of all recorded notifications of cleft palate.

- Among 1,434 infants with cleft palate and known outcome, 5.2% were stillborn; 8.4% of liveborn infants died in the neonatal period.

- Associated major malformations were reported in 19.7% of the births with cleft palate and another 7.5% had a chromosomal abnormality.

- For births in 1990 to 1994, the reported rates for cleft palate were highest in New South Wales (6.4 per 10,000 births) and lowest in the Australian Capital Territory (3.4 per 10,000 births) (Table 3.20, Figure 3.20).

Table 3.19: *Cleft palate by outcome and type of malformation, Australia, 1985-1994*

Outcome	1985	1986	1987	1988	1989	1990	1991	1992	1993	1994	1985-94
						Number					
Live births	106	130	118	128	143	154	147	153	152	129	1,360
Stillbirths	7	5	11	7	9	4	4	7	8	12	74
Total births*	113	135	129	135	152	160	153	161	161	142	1,441
Induced abortions	-	-	-	-	2	3	6	2	3	11	27
Neonatal deaths	10	17	16	13	16	9	7	8	6	12	114
						Rate per 10,000 births					
Total births	4.7	5.5	5.3	5.5	6.0	6.1	6.0	6.1	6.2	5.5	5.7
						Number					
Isolated	82	96	89	96	97	130	114	125	125	95	1,049
Associated	24	23	29	30	45	25	29	23	28	28	284
Chromosomal	7	16	11	9	10	5	10	13	8	19	108
						Rate per 10,000 births					
Isolated	3.4	3.9	3.6	3.9	3.8	4.9	4.4	4.8	4.8	3.7	4.1
Associated	1.0	0.9	1.2	1.2	1.8	0.9	1.1	0.9	1.1	1.1	1.1
Chromosomal	0.3	0.7	0.4	0.4	0.4	0.2	0.4	0.5	0.3	0.7	0.4

** Total includes 'not stated'*

Table 3.20: *Cleft palate, States and Territories, 1990-1994*

Outcome	NSW	Vic	Qld	WA	SA	Tas	ACT	NT	Australia
					Number				
Live births	271	187	124	62	58	17	7	9	735
Stillbirths	8	8	4	9	2	2	1	1	35
Total births*	282	196	128	73	61	19	8	10	777
Induced abortions	1	4	-	8	9	-	3	-	25
					Rate per 10,000 births				
Total births	6.4	6.0	5.5	5.8	6.1	5.5	3.4	5.6	6.0
					Number				
Isolated	213	158	102	46	41	14	6	9	589
Associated	48	25	18	20	17	4	-	1	133
Chromosomal	21	13	8	7	3	1	2	-	55
					Rate per 10,000 births				
Isolated	4.8	4.8	4.4	3.6	4.1	4.0	2.5	5.1	4.5
Associated	1.1	0.8	0.8	1.6	1.7	1.2	-	0.6	1.0
Chromosomal	0.5	0.4	0.3	0.6	0.3	0.3	0.8	-	0.4

** Total includes 'not stated'*

Figure 3.19: Cleft palate, Australia, 1985-1994

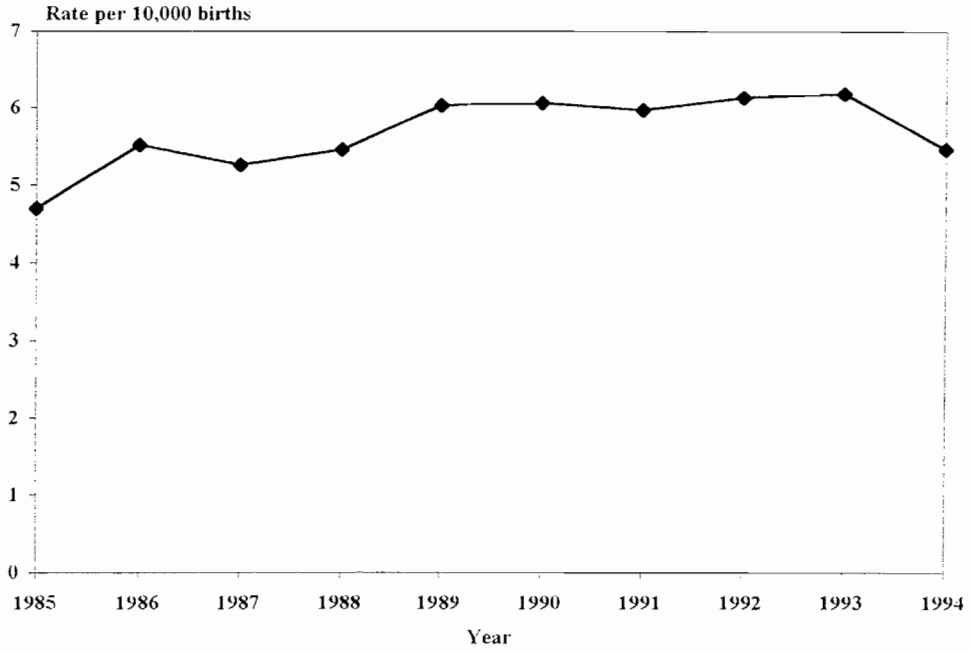
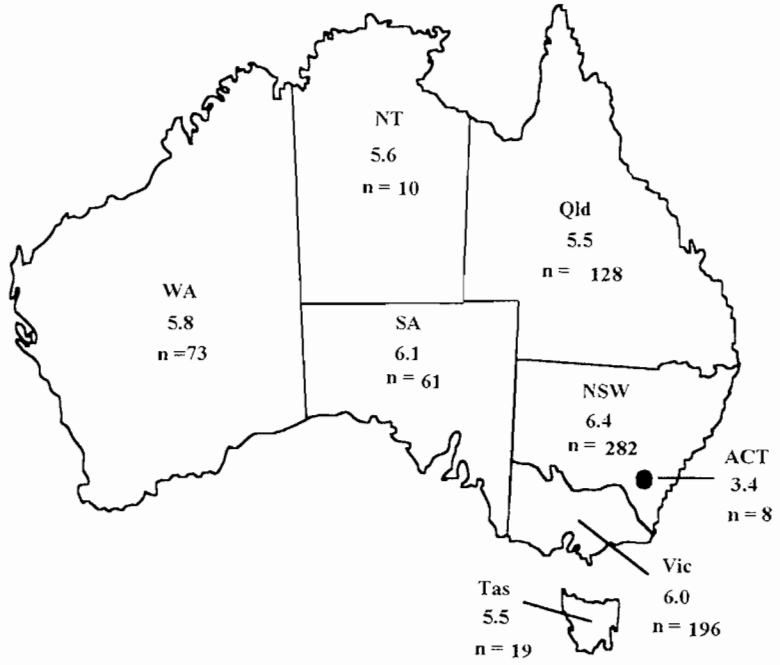


Figure 3.20: Cleft palate, States and Territories, 1990-1994

Australia: 6.0 per 10,000 births (n=777)



3.11 Cleft lip

- Cleft lip is a congenital malformation characterised by clefting of the upper lip, with or without clefting of the alveolar ridge and palate.
- The International Classification of Diseases codes are 749.1 for isolated cleft lip and 749.2 for cleft lip with cleft palate.
- The national rate of cleft lip in births showed very little change between 1985 and 1994, with an overall rate of 9.1 per 10,000 births in this period (Table 3.21, Figure 3.21).
- In the years 1985-1994, induced abortions were reported in 2.1% of all recorded notifications of cleft lip.
- Among 2,290 infants with cleft lip and known outcome, 6.3% were stillborn; 6.9% of liveborn infants died in the neonatal period.
- Associated major malformations were reported in 11.9% of the births with cleft lip and another 6.5% had a chromosomal abnormality.
- For births in 1990 to 1994, the reported rates for cleft lip were highest in Victoria (9.9 per 10,000 births) and lowest in the Australian Capital Territory and Western Australia (6.3 and 7.0 per 10,000 births, respectively) (Table 3.22, Figure 3.22).

Table 3.21: *Cleft lip with or without cleft palate by outcome and type of malformation, Australia, 1985-1994*

Outcome	1985	1986	1987	1988	1989	1990	1991	1992	1993	1994	1985-94
						Number					
Live births	202	207	225	194	239	231	198	203	221	226	2,146
Stillbirths	13	11	9	17	12	15	13	22	19	13	144
Total births*	215	218	234	211	252	248	212	225	241	240	2,296
Induced abortions	-	2	-	2	5	8	6	6	8	13	50
Neonatal deaths	19	19	20	9	26	7	17	13	9	9	148
						Rate per 10,000 births					
Total births	8.9	8.9	9.5	8.5	10.0	9.4	8.3	8.6	9.2	9.2	9.1
						Number					
Isolated	172	179	187	176	196	215	169	185	197	196	1,872
Associated	27	28	32	18	34	23	28	27	23	34	274
Chromosomal	16	11	15	17	22	10	15	13	21	10	150
						Rate per 10,000 births					
Isolated	7.1	7.3	7.6	7.1	7.8	8.1	6.6	7.0	7.6	7.5	7.4
Associated	1.1	1.1	1.3	0.7	1.3	0.9	1.1	1.0	0.9	1.3	1.1
Chromosomal	0.7	0.4	0.6	0.7	0.9	0.4	0.6	0.5	0.8	0.4	0.6

** Total includes 'not stated'*

Table 3.22: *Cleft lip with or without cleft palate, States and Territories, 1990-1994*

Outcome	NSW	Vic	Qld	WA	SA	Tas	ACT	NT	Australia
					Number				
Live births	359	299	206	78	83	24	14	16	1,079
Stillbirths	23	25	8	10	6	9	1	-	82
Total births*	386	324	214	88	90	33	15	16	1,166
Induced abortions	8	17	-	4	11	-	1	-	41
					Rate per 10,000 births				
Total births	8.7	9.9	9.3	7.0	9.0	9.5	6.3	9.0	8.9
					Number				
Isolated	312	269	181	69	77	27	14	13	962
Associated	48	41	18	14	9	1	1	3	135
Chromosomal	26	14	15	5	4	5	-	-	69
					Rate per 10,000 births				
Isolated	7.1	8.2	7.8	5.5	7.7	7.8	5.9	7.3	7.4
Associated	1.1	1.2	0.8	1.1	0.9	0.3	0.4	1.7	1.0
Chromosomal	0.6	0.4	0.6	0.4	0.4	1.4	-	-	0.5

** Total includes 'not stated'*

Figure 3.21: Cleft lip with or without cleft palate, Australia, 1985-1994

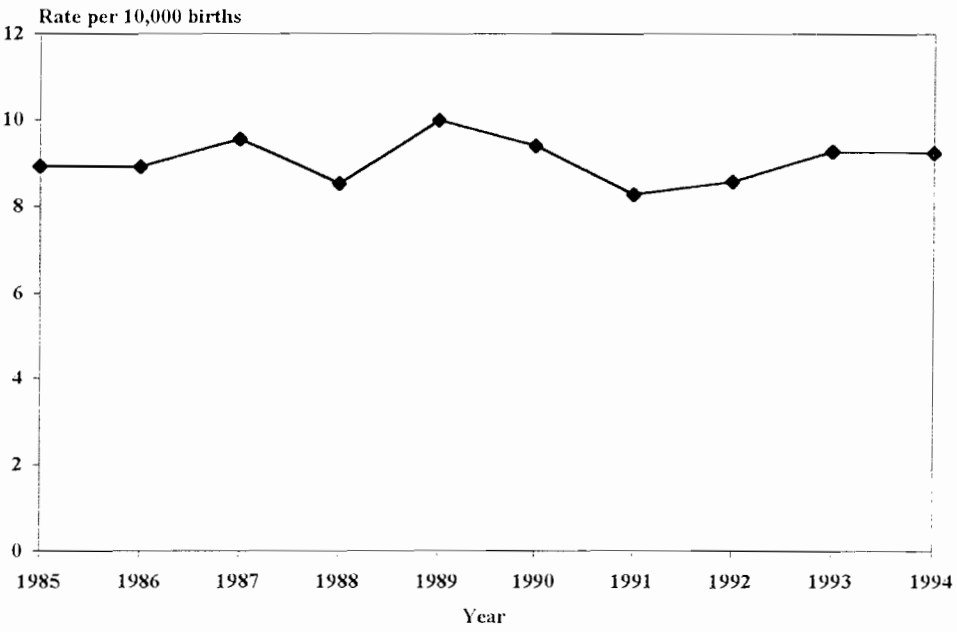
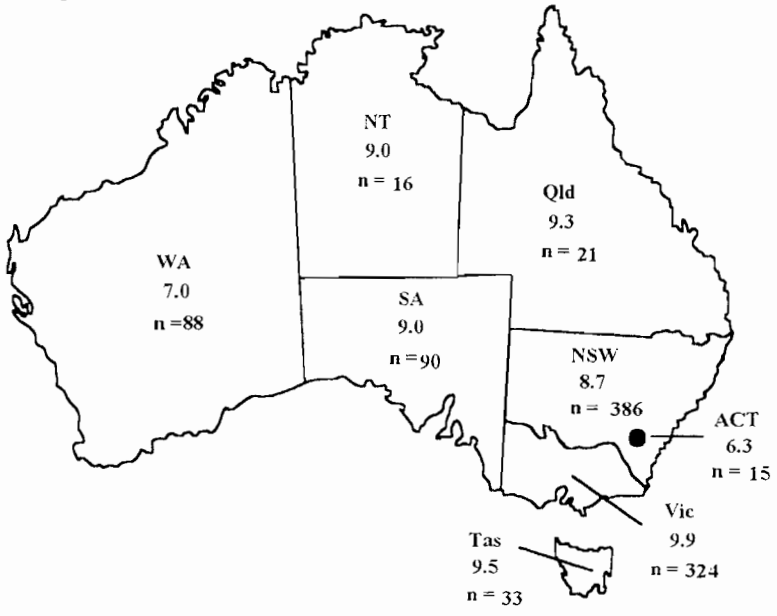


Figure 3.22: Cleft lip with or without cleft palate, States and Territories, 1990-1994

Australia: 8.9 per 10,000 births (n=1,166)



3.12 Oesophageal atresia or stenosis

- Oesophageal atresia or stenosis is a congenital malformation characterised by occlusion or narrowing of the oesophagus, with or without tracheo-oesophageal fistula.
- The International Classification of Diseases code for oesophageal atresia or stenosis is 750.3.
- The national rate of oesophageal atresia or stenosis in births was relatively stable between 1985 and 1994, with an overall rate of 3.1 per 10,000 births during this period (Table 3.23, Figure 3.23).
- In the years 1985-1994, induced abortions were reported in 0.5% of all recorded notifications of oesophageal atresia or stenosis.
- Among 776 infants with oesophageal atresia or stenosis and known outcome, 10.4% were stillborn; 15.7% of liveborn infants died in the neonatal period.
- Associated major malformations were reported in 42.1% of the births with oesophageal atresia or stenosis and another 8.8% had a chromosomal abnormality.
- For births in 1990 to 1994, the reported rates for oesophageal atresia or stenosis were highest in South Australia (5.0 per 10,000 births) and lowest in the Australian Capital Territory and the Northern Territory (1.3 and 1.7 per 10,000 births, respectively) (Table 3.24, Figure 3.24).

Table 3.23: *Oesophageal atresia/stenosis by outcome and type of malformation, Australia, 1985-1994*

Outcome	1985	1986	1987	1988	1989	1990	1991	1992	1993	1994	1985-94
						Number					
Live births	65	53	79	62	65	73	71	79	89	59	695
Stillbirths	12	5	9	7	8	9	6	9	9	7	81
Total births*	77	58	88	69	73	82	77	88	98	66	776
Induced abortions	-	-	-	-	-	-	1	-	2	1	4
Neonatal deaths	13	12	14	15	6	16	10	6	15	2	109
						Rate per 10,000 births					
Total births	3.2	2.4	3.6	2.8	2.9	3.1	3.0	3.3	3.8	2.5	3.1
						Number					
Isolated	38	30	45	38	34	43	44	45	37	27	381
Associated	36	19	32	26	30	31	27	39	53	34	327
Chromosomal	3	9	11	5	9	8	6	4	8	5	68
						Rate per 10,000 births					
Isolated	1.6	1.2	1.8	1.5	1.3	1.6	1.7	1.7	1.4	1.0	1.5
Associated	1.5	0.8	1.3	1.0	1.2	1.2	1.1	1.5	2.0	1.3	1.3
Chromosomal	0.1	0.4	0.4	0.2	0.4	0.3	0.2	0.2	0.3	0.2	0.3

** Total includes 'not stated'*

Table 3.24: *Oesophageal atresia/stenosis, States and Territories, 1990-1994*

Outcome	NSW	Vic	Qld	WA	SA	Tas	ACT	NT	Australia
					Number				
Live births	120	103	72	22	41	8	2	3	371
Stillbirths	10	11	1	4	9	4	1	-	40
Total births*	130	114	73	26	50	12	3	3	411
Induced abortions	-	2	-	2	-	-	-	-	4
					Rate per 10,000 births				
Total births	2.9	3.5	3.2	2.1	5.0	3.5	1.3	1.7	3.2
					Number				
Isolated	54	62	44	9	16	7	2	2	196
Associated	64	44	27	13	30	4	1	1	184
Chromosomal	12	8	2	4	4	1	-	-	31
					Rate per 10,000 births				
Isolated	1.2	1.9	1.9	0.7	1.6	2.0	0.8	1.1	1.5
Associated	1.4	1.3	1.2	1.0	3.0	1.2	0.4	0.6	1.4
Chromosomal	0.3	0.2	0.1	0.3	0.4	0.3	-	-	0.2

** Total includes 'not stated'*

Figure 3.23: Oesophageal atresia/stenosis, Australia, 1985-1994

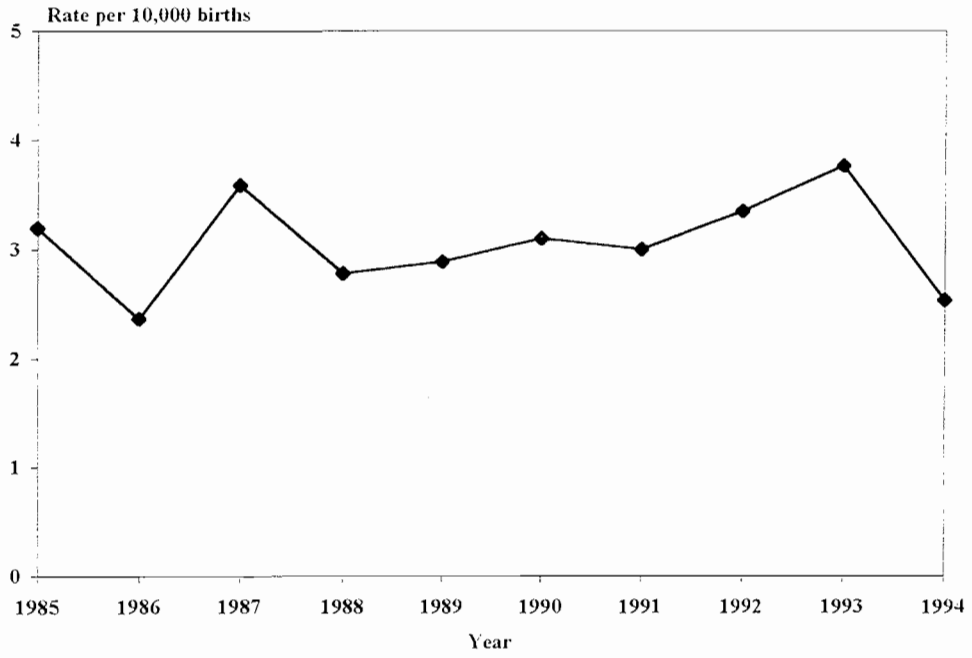
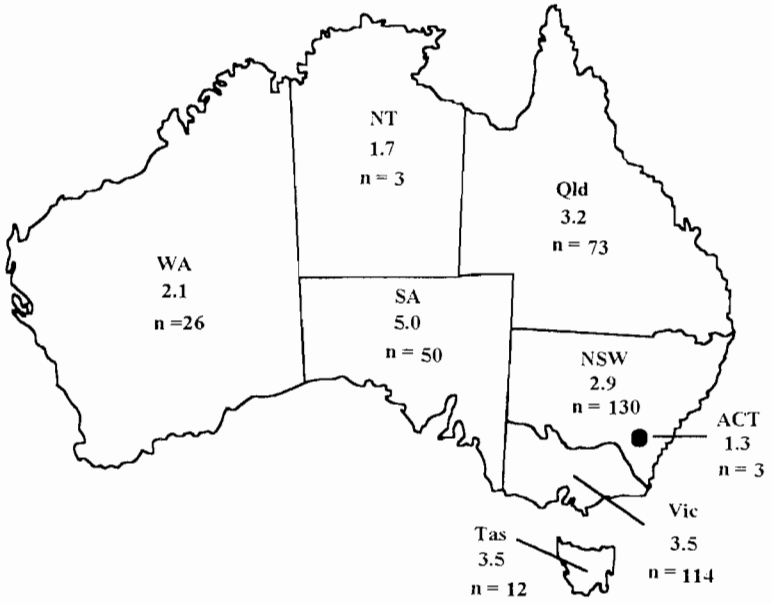


Figure 3.24: Oesophageal atresia/stenosis, States and Territories, 1990-1994

Australia: 3.2 per 10,000 births (n=411)



3.13 Small intestinal atresia or stenosis

- Small intestinal atresia or stenosis is a congenital malformation characterised by occlusion or narrowing of the duodenum, jejunum or ileum.

- The International Classification of Diseases code for small intestinal atresia or stenosis is 751.1.

- The national rate of small intestinal atresia or stenosis in births was relatively stable between 1985 and 1994, with an overall rate of 2.1 per 10,000 births during this period (Table 3.25, Figure 3.25).

- Among 535 infants with small intestinal atresia or stenosis and known outcome, 5.8% were stillborn; 12.3% of liveborn infants died in the neonatal period.

- Associated major malformations were reported in 22.2% of the births with small intestinal atresia or stenosis and another 19.6% had a chromosomal abnormality.

- For births in 1990 to 1994, the reported rates for small intestinal atresia or stenosis were highest in South Australia (3.3 per 10,000 births) and lowest in the Australian Capital Territory, the Northern Territory and Tasmania (0.8, 1.1 and 1.2 per 10,000 births, respectively) (Table 3.26, Figure 3.26).

Table 3.25: *Small intestinal atresia/stenosis by outcome and type of malformation, Australia, 1985-1994*

Outcome	1985	1986	1987	1988	1989	1990	1991	1992	1993	1994	1985-94
						Number					
Live births	53	42	46	62	42	54	58	57	50	40	504
Stillbirths	3	2	3	2	3	4	2	2	3	7	31
Total births*	56	44	49	64	45	58	60	60	53	48	537
Induced abortions	-	-	-	-	-	-	-	-	-	1	1
Neonatal deaths	13	7	8	8	8	4	8	1	3	2	62
						Rate per 10,000 births					
Total births	2.3	1.8	2.0	2.6	1.8	2.2	2.3	2.3	2.0	1.8	2.1
						Number					
Isolated	37	32	22	36	20	36	31	40	33	26	313
Associated	11	4	14	13	12	15	19	13	9	9	119
Chromosomal	8	8	13	15	13	7	10	7	11	13	105
						Rate per 10,000 births					
Isolated	1.5	1.3	0.9	1.5	0.8	1.4	1.2	1.5	1.3	1.0	1.2
Associated	0.5	0.2	0.6	0.5	0.5	0.6	0.7	0.5	0.3	0.3	0.5
Chromosomal	0.3	0.3	0.5	0.6	0.5	0.3	0.4	0.3	0.4	0.5	0.4

* *Total includes 'not stated'*

Table 3.26: *Small intestinal atresia/stenosis, States and Territories, 1990-1994*

Outcome	NSW	Vic	Qld	WA	SA	Tas	ACT	NT	Australia
					Number				
Live births	77	74	47	23	31	3	2	2	259
Stillbirths	5	3	3	5	1	1	-	-	18
Total births*	83	77	50	28	33	4	2	2	279
Induced abortions	-	-	-	-	1	-	-	-	1
					Rate per 10,000 births				
Total births	1.9	2.3	2.2	2.2	3.3	1.2	0.8	1.1	2.1
					Number				
Isolated	48	43	34	17	19	4	1	-	166
Associated	21	18	9	8	8	-	-	1	65
Chromosomal	14	16	7	3	6	-	1	1	48
					Rate per 10,000 births				
Isolated	1.1	1.3	1.5	1.3	1.9	1.2	0.4	-	1.3
Associated	0.5	0.5	0.4	0.6	0.8	-	-	0.6	0.5
Chromosomal	0.3	0.5	0.3	0.2	0.6	-	0.4	0.6	0.4

* *Total includes 'not stated'*

Figure 3.25: Small intestinal atresia/stenosis, Australia, 1985-1994

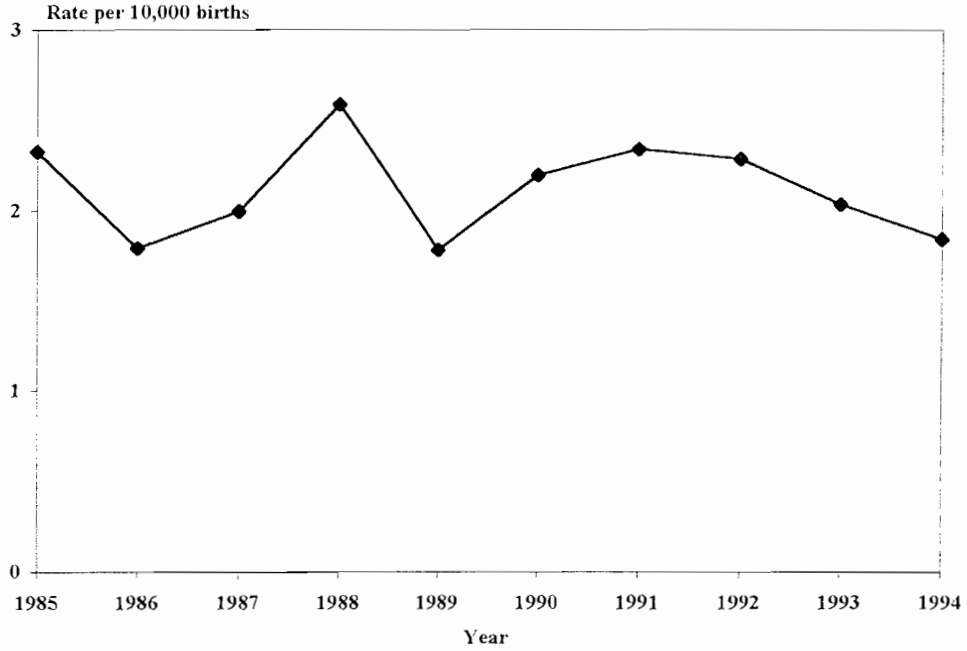
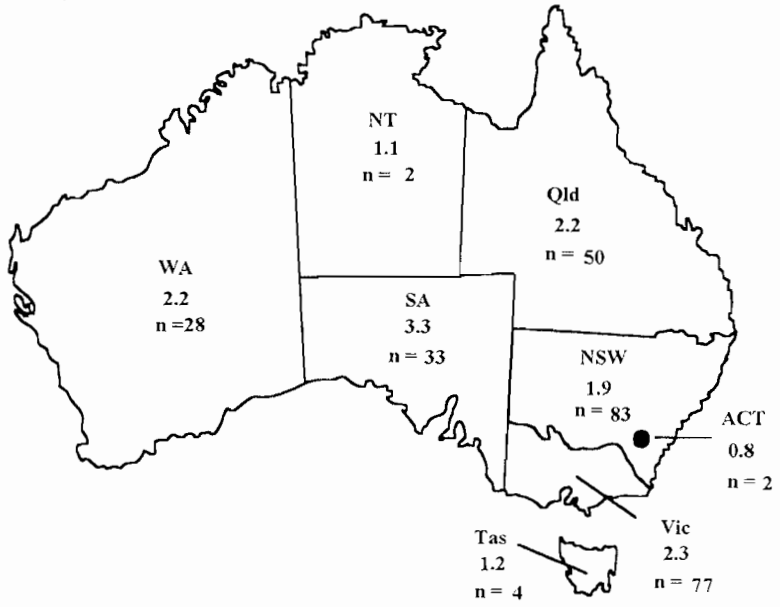


Figure 3.26: Small intestinal atresia/stenosis, States and Territories, 1990-1994

Australia: 2.1 per 10,000 births (n=279)



3.14 Anorectal atresia or stenosis

- Anorectal atresia or stenosis is a congenital malformation characterised by absence of the anus or of the communication between rectum and anus, or narrowing of the canal, with or without fistula to neighbouring organs. Clinically, these malformations are often termed imperforate anus.

- The International Classification of Diseases code for anorectal atresia or stenosis is 751.2. The British Paediatric Association Classification codes for atresia or stenosis of the rectum or anus are 751.21-751.24.

- The national rate of anorectal atresia or stenosis in births has varied little since 1988, with an overall rate of 3.3 per 10,000 births during 1985 to 1994 (Table 3.27, Figure 3.27).

- In the years 1985-1994, induced abortions were reported in 4.2% of all recorded notifications of anorectal atresia or stenosis.

- Among 825 infants with anorectal atresia or stenosis and known outcome, 12.2% were stillborn; 19.1% of liveborn infants died in the neonatal period.

- Associated major malformations were reported in 53.9% of the births with anorectal atresia or stenosis and another 6.7% had a chromosomal abnormality.

- For births in 1990 to 1994, the reported rates for anorectal atresia or stenosis were highest in South Australia and the Northern Territory (4.2 and 3.9 per 10,000 births, respectively) and lowest in Tasmania and the Australian Capital Territory (0.3 and 0.4 per 10,000 births, respectively) (Table 3.28, Figure 3.28). There was only 1 reported birth each in Tasmania and the Australian Capital Territory.

Table 3.27: *Atresia/stenosis of large intestine, rectum or anal canal by outcome and type of malformation Australia, 1985-1994*

Outcome	1985	1986	1987	1988	1989	1990	1991	1992	1993	1994	1985-94
						Number					
Live births	69	60	89	67	66	73	74	75	79	72	724
Stillbirths	14	12	10	7	11	14	13	9	6	5	101
Total births*	83	72	99	74	77	87	87	84	85	77	825
Induced abortions	-	2	-	2	1	5	5	1	8	12	36
Neonatal deaths	24	15	22	20	10	15	12	4	8	8	138
						Rate per 10,000 births					
Total births	3.4	2.9	4.0	3.0	3.1	3.3	3.4	3.2	3.3	3.0	3.3
						Number					
Isolated	27	30	35	30	26	34	32	39	39	33	325
Associated	53	37	56	41	44	49	50	37	41	37	445
Chromosomal	3	5	8	3	7	4	5	8	5	7	55
						Rate per 10,000 births					
Isolated	1.1	1.2	1.4	1.2	1.0	1.3	1.2	1.5	1.5	1.3	1.3
Associated	2.2	1.5	2.3	1.7	1.7	1.9	1.9	1.4	1.6	1.4	1.8
Chromosomal	0.1	0.2	0.3	0.1	0.3	0.2	0.2	0.3	0.2	0.3	0.2

** Total includes 'not stated'*

Table 3.28: *Atresia/stenosis of large intestine, rectum or anal canal, States and Territories, 1990-1994*

Outcome	NSW	Vic	Qld	WA	SA	Tas	ACT	NT	Australia
					Number				
Live births	115	111	67	37	35	1	-	7	373
Stillbirths	12	17	5	5	7	-	1	-	47
Total births*	127	128	72	42	42	1	1	7	420
Induced abortions	5	8	-	8	10	-	-	-	31
					Rate per 10,000 births				
Total births	2.9	3.9	3.1	3.3	4.2	0.3	0.4	3.9	3.2
					Number				
Isolated	44	61	40	13	16	1	-	2	177
Associated	76	59	30	23	21	-	1	4	214
Chromosomal	7	8	2	6	5	-	-	1	29
					Rate per 10,000 births				
Isolated	1.0	1.9	1.7	1.0	1.6	0.3	-	1.1	1.4
Associated	1.7	1.8	1.3	1.8	2.1	-	0.4	2.3	1.6
Chromosomal	0.2	0.2	0.1	0.5	0.5	-	-	0.6	0.2

** Total includes 'not stated'*

Figure 3.27: Atresia/stenosis of large intestine, rectum or anal canal, Australia, 1985-1994

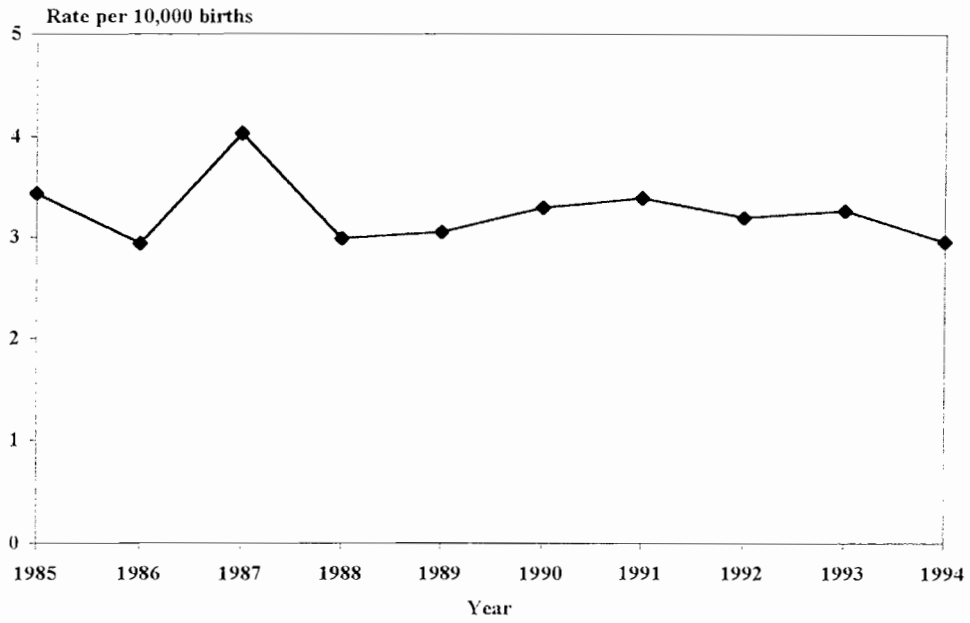
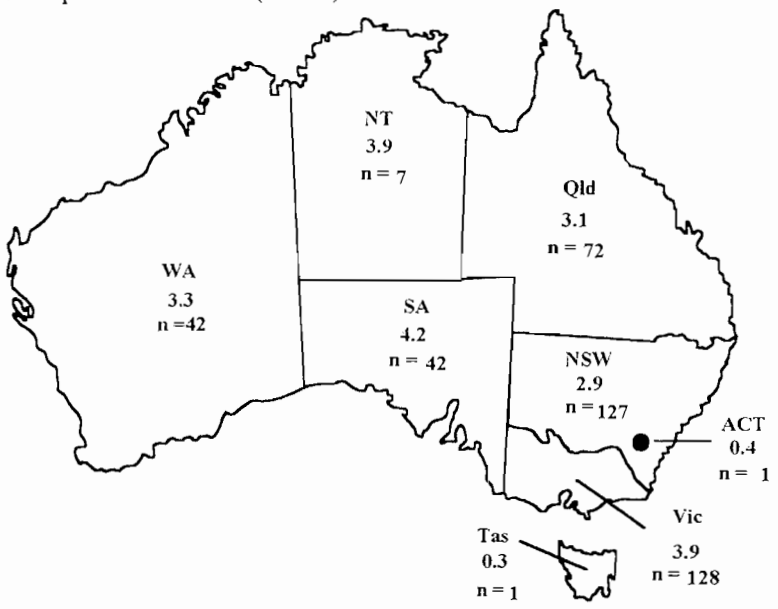


Figure 3.28: Atresia/stenosis of large intestine, rectum or anal canal, States and Territories, 1990-1994

Australia: 3.2 per 10,000 births (n=420)



3.15 Hypospadias

- Hypospadias is a congenital malformation characterised by opening of the urethra on the ventral side of the penis, irrespective of the degree of severity.

- The International Classification of Diseases code for hypospadias is 752.6. Although epispadias and congenital chordee are also included under this code in ICD, separate 5-digit codes in the British Paediatric Association Classification classification enable distinction between hypospadias and these other malformations.

- The national rate of hypospadias in births increased each year from 15.1 per 10,000 births in 1985 to 23.1 per 10,000 births in 1991 and then decreased each year to 20.5 per 10,000 births in 1994 (Table 3.29, Figure 3.29).

- Among 5,093 infants with hypospadias and known outcome, 0.8% were stillborn; 1.8% of liveborn infants died in the neonatal period.

- Associated major malformations were reported in 6.3% of the births with hypospadias and another 1.4% had a chromosomal abnormality.

- For births in 1990 to 1994, the reported rates for hypospadias were highest in Victoria (27.6 per 10,000 births) and lowest in the Northern Territory (3.4 per 10,000 births) (Table 3.30, Figure 3.30).

Table 3.29: *Hypospadias by outcome and type of malformation, Australia, 1985-1994*

Outcome	1985	1986	1987	1988	1989	1990	1991	1992	1993	1994	1985-94
						Number					
Live births	361	432	451	477	513	561	591	585	548	533	5,052
Stillbirths	4	3	2	4	4	6	1	13	2	2	41
Total births*	365	435	453	481	517	567	593	600	550	537	5,098
Induced abortions	-	-	-	-	1	-	-	1	4	2	8
Neonatal deaths	13	9	9	10	12	11	6	9	5	8	92
						Rate per 10,000 births					
Total births	15.1	17.8	18.5	19.4	20.5	21.5	23.1	22.8	21.1	20.5	20.1
						Number					
Isolated	329	405	402	452	470	524	559	571	508	484	4,704
Associated	26	24	45	23	36	38	27	26	33	45	323
Chromosomal	10	6	6	6	11	5	7	3	9	8	71
						Rate per 10,000 births					
Isolated	13.6	16.5	16.4	18.3	18.6	19.8	21.8	21.7	19.5	18.5	18.5
Associated	1.1	1.0	1.8	0.9	1.4	1.4	1.1	1.0	1.3	1.7	1.3
Chromosomal	0.4	0.2	0.2	0.2	0.4	0.2	0.3	0.1	0.3	0.3	0.3

** Total includes 'not stated'*

Table 3.30: *Hypospadias, States and Territories, 1990-1994*

Outcome	NSW	Vic	Qld	WA	SA	Tas	ACT	NT	Australia
					Number				
Live births	894	905	473	234	236	36	34	6	2,818
Stillbirths	6	3	1	1	1	12	-	-	24
Total births*	901	908	478	235	237	48	34	6	2,847
Induced abortions	1	2	-	1	3	-	-	-	7
					Rate per 10,000 births				
Total births	20.4	27.6	20.6	18.6	23.8	13.8	14.3	3.4	21.8
					Number				
Isolated	820	856	451	215	220	48	32	4	2,646
Associated	69	42	23	17	14	-	2	2	169
Chromosomal	12	10	4	3	3	-	-	-	32
					Rate per 10,000 births				
Isolated	18.5	26.0	19.5	17.0	22.1	13.8	13.5	2.3	20.3
Associated	1.6	1.3	1.0	1.3	1.4	-	0.8	1.1	1.3
Chromosomal	0.3	0.3	0.2	0.2	0.3	-	-	-	0.2

** Total includes 'not stated'*

Figure 3.29: Hypospadias, Australia, 1985-1994

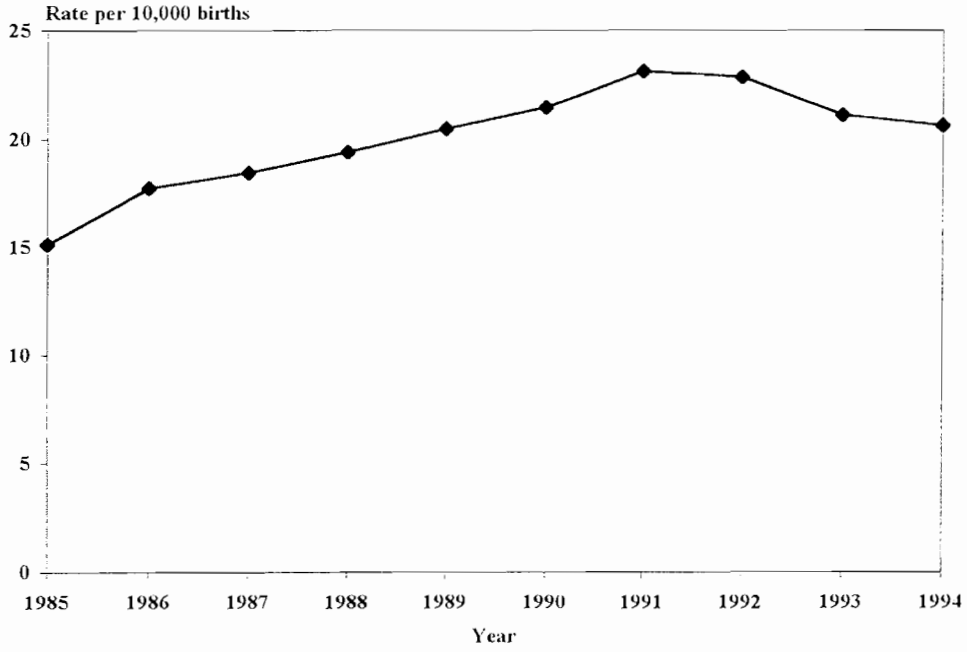
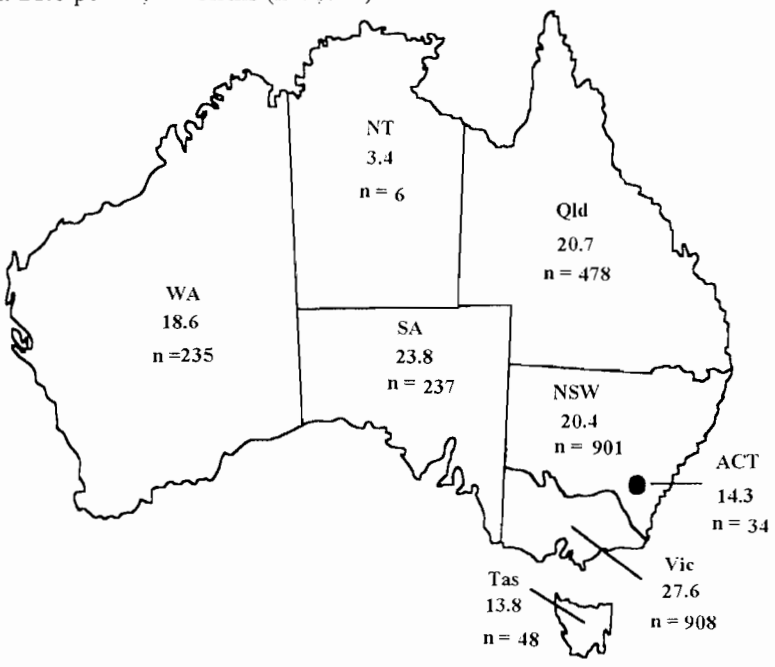


Figure 3.30: Hypospadias, States and Territories, 1990-1994

Australia: 21.8 per 10,000 births (n=2,847)



3.16 Renal agenesis and dysgenesis

- Renal agenesis and dysgenesis are congenital malformations characterised by absence of kidneys or severely dysplastic kidneys and occurring bilaterally or unilaterally. Polycystic kidneys are not included in this group.

- The International Classification of Diseases code for renal agenesis and dysgenesis is 753.0.

- The national rate of renal agenesis and dysgenesis in births varied between high rates of 4.1 and 4.2 per 10,000 births (in 1985 and 1991, respectively) and low rates of 2.7 and 2.6 per 10,000 births (in 1986 and 1992, respectively) (Table 3.31, Figure 3.31). There were more induced abortions performed before 20 weeks' gestation for renal agenesis and dysgenesis in 1993 and 1994 than in previous years.

- In the years 1985-1994, induced abortions were reported in 7.2% of all recorded notifications of renal agenesis and dysgenesis.

- Among 858 infants with renal agenesis and dysgenesis and known outcome, 27.3% were stillborn; 50.8% of liveborn infants died in the neonatal period. Almost two-thirds of infants have bilateral agenesis and dysgenesis (Table 5), which is a lethal malformation.

- Associated major malformations were reported in 46.7% of the births with renal agenesis and dysgenesis and another 5.5% had a chromosomal abnormality.

- For births in 1990 to 1994, the reported rates for renal agenesis and dysgenesis were highest in South Australia and Queensland (4.7 and 4.0 per 10,000 births, respectively) and lowest in the Australian Capital Territory (0.8 per 10,000 births) (Table 3.32, Figure 3.32).

Table 3.31: *Renal agenesis/dysgenesis by outcome and type of malformation, Australia, 1985-1994*

Outcome	1985	1986	1987	1988	1989	1990	1991	1992	1993	1994	1985-94
						Number					
Live births	72	46	69	61	59	73	77	49	52	66	624
Stillbirths	28	20	20	19	33	24	32	20	20	18	234
Total births*	100	66	90	81	92	97	109	69	73	84	861
Induced abortions	2	2	-	8	8	4	3	6	15	19	67
Neonatal deaths	56	35	47	38	30	35	37	9	15	15	317
						Rate per 10,000 births					
Total births	4.1	2.7	3.7	3.3	3.6	3.7	4.2	2.6	2.8	3.2	3.4
						Number					
Isolated	41	25	45	35	43	48	60	41	36	38	412
Associated	53	38	40	42	44	43	44	27	33	38	402
Chromosomal	6	3	5	4	5	6	5	1	4	8	47
						Rate per 10,000 births					
Isolated	1.7	1.0	1.8	1.4	1.7	1.8	2.3	1.6	1.4	1.5	1.6
Associated	2.2	1.6	1.6	1.7	1.7	1.6	1.7	1.0	1.3	1.5	1.6
Chromosomal	0.2	0.1	0.2	0.2	0.2	0.2	0.2	0.0	0.2	0.3	0.2

* *Total includes 'not stated'*

Table 3.32: *Renal agenesis/dysgenesis, States and Territories, 1990-1994*

Outcome	NSW	Vic	Qld	WA	SA	Tas	ACT	NT	Australia
					Number				
Live births	88	90	72	25	30	5	2	5	317
Stillbirths	30	34	21	11	16	1	-	1	114
Total births*	118	124	93	36	47	6	2	6	432
Induced abortions	11	14	-	9	12	1	-	-	47
					Rate per 10,000 births				
Total births	2.7	3.8	4.0	2.8	4.7	1.7	0.8	3.4	3.3
					Number				
Isolated	54	60	63	15	21	4	1	5	223
Associated	60	53	25	19	24	2	1	1	185
Chromosomal	4	11	5	2	2	-	-	-	24
					Rate per 10,000 births				
Isolated	1.2	1.8	2.7	1.2	2.1	1.2	0.4	2.8	1.7
Associated	1.4	1.6	1.1	1.5	2.4	0.6	0.4	0.6	1.4
Chromosomal	0.1	0.3	0.2	0.2	0.2	-	-	-	0.2

* *Total includes 'not stated'*

Figure 3.31: Renal agenesis/ dysgenesis, Australia, 1985-1994

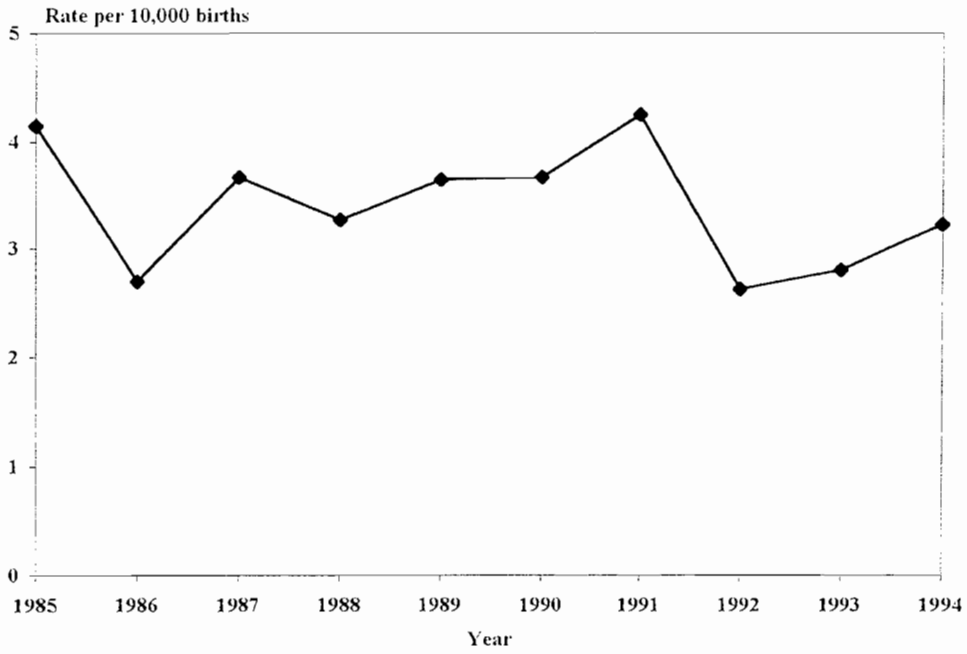
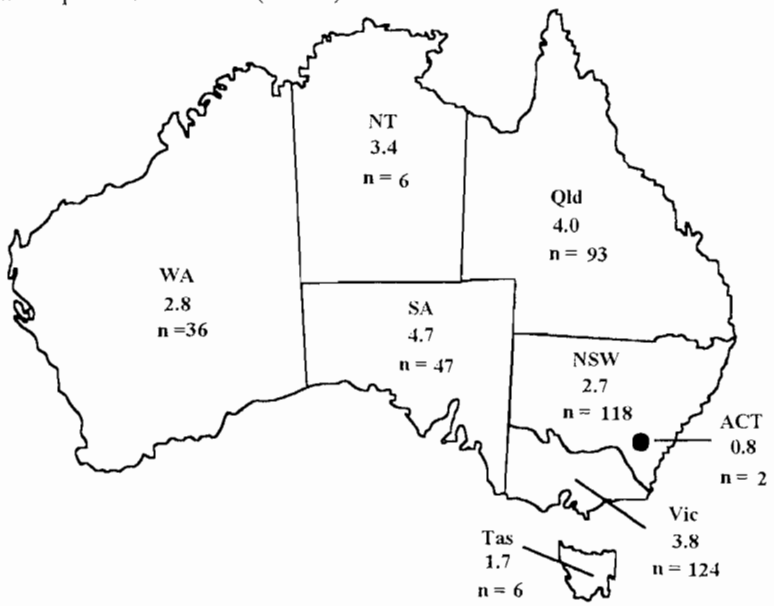


Figure 3.32: Renal agenesis/dysgenesis, States and Territories, 1990-1994

Australia: 3.3 per 10,000 births (n=432)



3.17 Cystic kidney disease

- Cystic kidney disease includes a heterogeneous group of malformations characterised by renal cysts of variable size and extent and described as polycystic or multicystic kidneys, occurring bilaterally or unilaterally.

- The International Classification of Diseases code for cystic kidney disease is 753.1. The British Paediatric Association Classification enables distinction between the various pathological types.

- The national rate of cystic kidney disease in births was higher in the 1990s than in the late 1980s, with a trough in 1987 of 2.0 per 10,000 births and a peak of 3.5 per 10,000 births in 1991 (Table 3.33, Figure 3.33).

- In the years 1985-1994, induced abortions were reported in 7.3% of all recorded notifications of cystic kidney disease.

- Among 679 infants with cystic kidney disease and known outcome, 13.5% were stillborn; 32.5% of liveborn infants died in the neonatal period.

- Associated major malformations were reported in 34.2% of the births with cystic kidney disease and another 7.0% had a chromosomal abnormality.

- For births in 1990 to 1994, the reported rates for cystic kidney disease were highest in the Northern Territory (6.8 per 10,000 births) and lowest in the Australian Capital Territory and Tasmania (0.8 and 1.4 per 10,000 births, respectively) (Table 3.34, Figure 3.34).

Table 3.33: *Cystic kidney disease by outcome and type of malformation, Australia, 1985-1994*

Outcome	1985	1986	1987	1988	1989	1990	1991	1992	1993	1994	1985-94
						Number					
Live births	49	44	46	47	54	73	77	67	63	67	587
Stillbirths	13	10	3	9	11	5	12	9	8	12	92
Total births*	62	54	49	56	65	79	89	77	74	79	684
Induced abortions	-	3	4	4	2	4	3	9	7	18	54
Neonatal deaths	29	23	23	19	19	24	22	11	8	13	191
						Rate per 10,000 births					
Total births	2.6	2.2	2.0	2.3	2.6	3.0	3.5	2.9	2.8	3.0	2.7
						Number					
Isolated	28	31	20	29	37	48	57	57	53	42	402
Associated	30	20	24	21	20	26	29	18	14	32	234
Chromosomal	4	3	5	6	8	5	3	2	7	5	48
						Rate per 10,000 births					
Isolated	1.2	1.3	0.8	1.2	1.5	1.8	2.2	2.2	2.0	1.6	1.6
Associated	1.2	0.8	1.0	0.8	0.8	1.0	1.1	0.7	0.5	1.2	0.9
Chromosomal	0.2	0.1	0.2	0.2	0.3	0.2	0.1	0.1	0.3	0.2	0.2

* *Total includes 'not stated'*

Table 3.34: *Cystic kidney disease, States and Territories, 1990-1994*

Outcome	NSW	Vic	Qld	WA	SA	Tas	ACT	NT	Australia
					Number				
Live births	93	105	65	28	39	5	2	10	347
Stillbirths	13	16	4	6	5	-	-	2	46
Total births*	109	121	69	35	45	5	2	12	398
Induced abortions	1	13	-	15	11	1	-	-	41
					Rate per 10,000 births				
Total births	2.5	3.7	3.0	2.8	4.5	1.4	0.8	6.8	3.1
					Number				
Isolated	71	85	40	16	32	4	2	7	257
Associated	32	32	24	14	11	1	-	5	119
Chromosomal	6	4	5	5	2	-	-	-	22
					Rate per 10,000 births				
Isolated	1.6	2.6	1.7	1.3	3.2	1.2	0.8	3.9	2.0
Associated	0.7	1.0	1.0	1.1	1.1	0.3	-	2.8	0.9
Chromosomal	0.1	0.1	0.2	0.4	0.2	-	-	-	0.2

* *Total includes 'not stated'*

Figure 3.33: Cystic kidney disease, Australia, 1985-1994

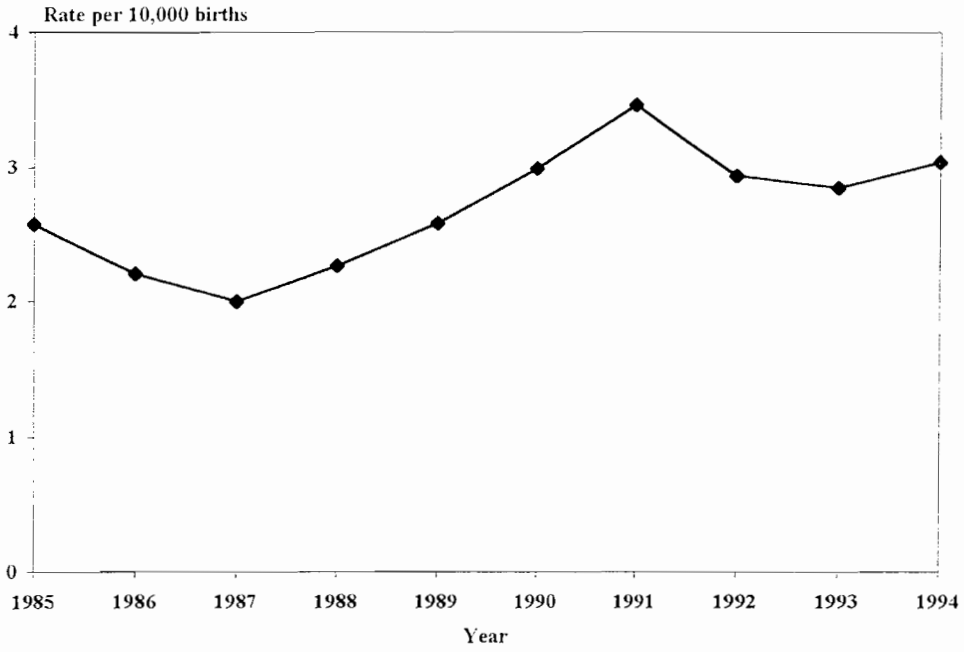
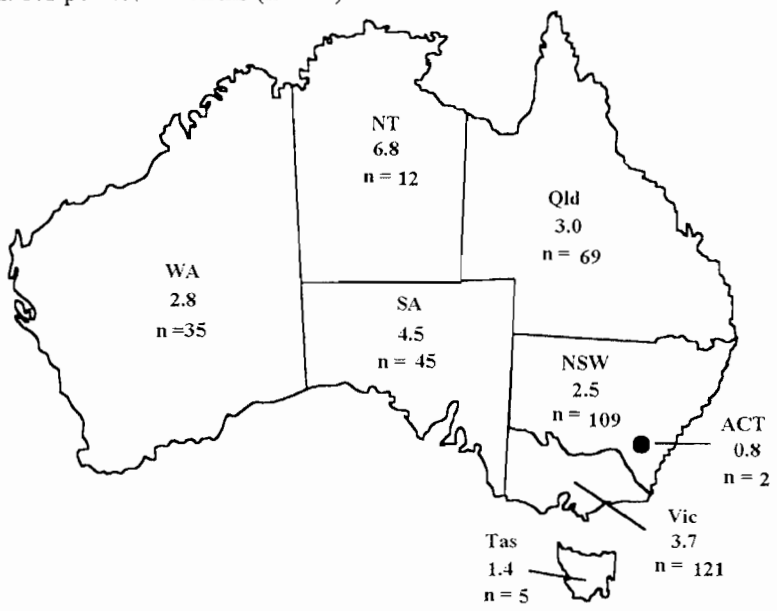


Figure 3.34: Cystic kidney disease, States and Territories, 1990-1994

Australia: 3.1 per 10,000 births (n=398)



3.18 Obstructive defects of renal pelvis and ureter

- This group of malformations includes hydronephrosis and other obstructive defects of the renal pelvis and ureter that result in dilation of the renal collecting systems, occurring bilaterally or unilaterally.

- The International Classification of Diseases code for obstructive defects of renal pelvis and ureter is 753.2.

- The national rate of obstructive defects of renal pelvis and ureter in births has more than tripled from 3.4 per 10,000 births in 1985 to 10.5 per 10,000 births in 1994 (Table 3.35, Figure 3.35). These malformations are increasingly detected by prenatal ultrasound screening.

- In the years 1985-1994, induced abortions were reported in 2.0% of all recorded notifications of obstructive defects of renal pelvis and ureter.

- Among 1,583 infants with obstructive defects of renal pelvis and ureter and known outcome, 5.7% were stillborn; 8.8% of liveborn infants died in the neonatal period.

- Associated major malformations were reported in 23.2% of the births with obstructive defects of renal pelvis and ureter and another 5.9% had a chromosomal abnormality.

- For births in 1990 to 1994, the reported rates for obstructive defects of renal pelvis and ureter were highest in Victoria (13.8 per 10,000 births) and lowest in Tasmania (2.3 per 10,000 births) (Table 3.36, Figure 3.36).

Table 3.35: *Obstructive defects of renal pelvis and ureter by outcome and type of malformation, Australia, 1985-1994*

Outcome	1985	1986	1987	1988	1989	1990	1991	1992	1993	1994	1985-94
						Number					
Live births	70	84	107	100	135	184	185	181	186	261	1,493
Stillbirths	11	8	9	7	6	7	9	9	12	12	90
Total births*	81	92	116	107	141	193	196	190	201	275	1,592
Induced abortions	-	1	1	-	5	6	-	3	10	7	33
Neonatal deaths	19	20	16	15	10	9	10	11	11	10	131
						Rate per 10,000 births					
Total births	3.4	3.8	4.7	4.3	5.6	7.3	7.6	7.2	7.7	10.5	6.3
						Number					
Isolated	40	46	67	74	95	138	148	152	153	215	1,128
Associated	32	35	40	26	36	46	39	29	42	45	370
Chromosomal	9	11	9	7	10	9	9	9	6	15	94
						Rate per 10,000 births					
Isolated	1.7	1.9	2.7	3.0	3.8	5.2	5.8	5.8	5.9	8.2	4.4
Associated	1.3	1.4	1.6	1.0	1.4	1.7	1.5	1.1	1.6	1.7	1.5
Chromosomal	0.4	0.4	0.4	0.3	0.4	0.3	0.4	0.3	0.2	0.6	0.4

** Total includes 'not stated'*

Table 3.36: *Obstructive defects of renal pelvis and ureter, States and Territories, 1990-1994*

Outcome	NSW	Vic	Qld	WA	SA	Tas	ACT	NT	Australia
					Number				
Live births	278	435	135	39	73	7	15	15	997
Stillbirths	8	14	2	10	7	1	5	2	49
Total births*	290	452	139	49	80	8	20	17	1,055
Induced abortions	3	13	-	5	5	-	-	-	26
					Rate per 10,000 births				
Total births	6.6	13.8	6.0	3.9	8.0	2.3	8.4	9.6	8.1
					Number				
Isolated	205	375	117	21	53	8	16	11	806
Associated	73	59	19	20	21	-	3	6	201
Chromosomal	12	18	3	8	6	-	1	-	48
					Rate per 10,000 births				
Isolated	4.6	11.4	5.1	1.7	5.3	2.3	6.7	6.2	6.2
Associated	1.6	1.8	0.8	1.6	2.1	-	1.3	3.4	1.5
Chromosomal	0.3	0.5	0.1	0.6	0.6	-	0.4	-	0.4

** Total includes 'not stated'*

Figure 3.35: Obstructive defects of renal pelvis and ureter, Australia, 1985-1994

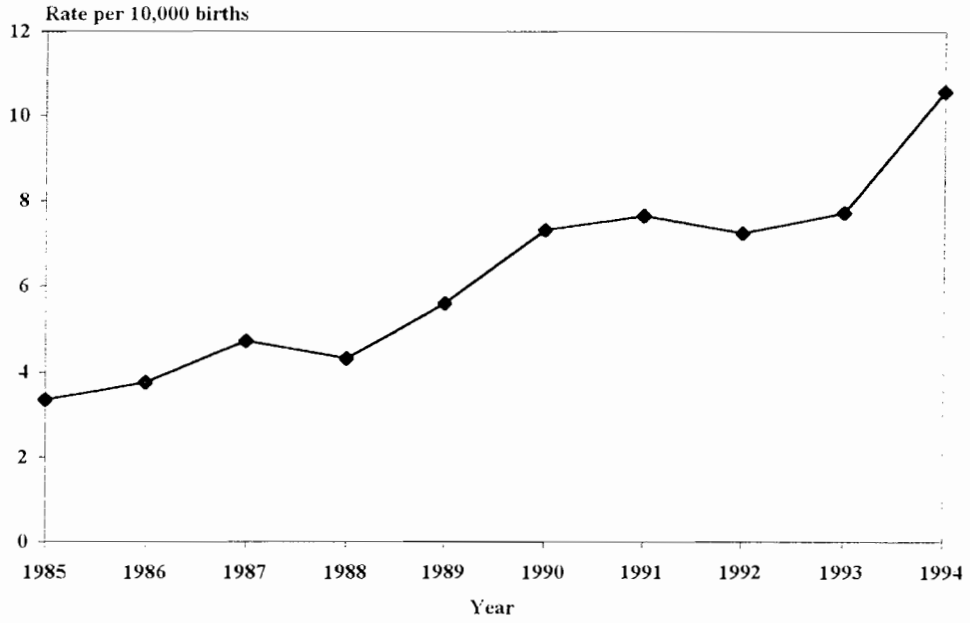
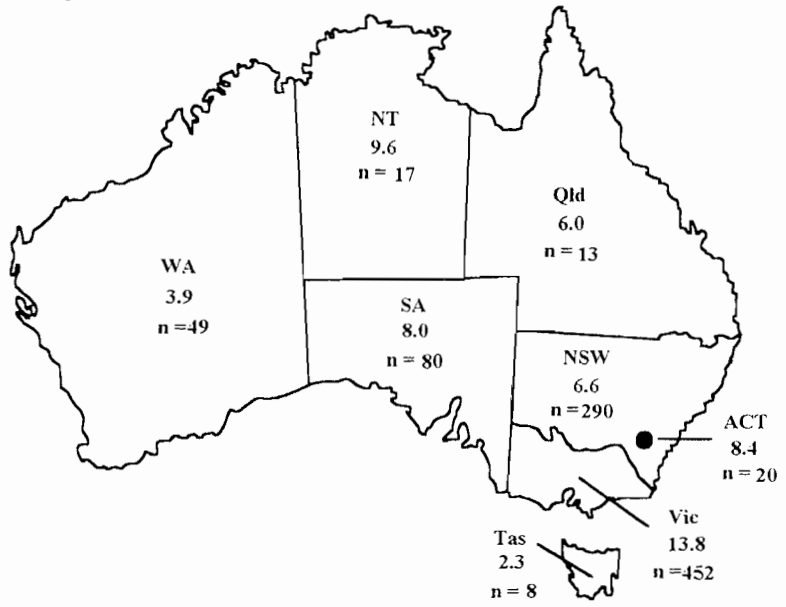


Figure 3.36: Obstructive defects of renal pelvis and ureter, States and Territories, 1990-1994

Australia: 8.1 per 10,000 births (n=1,055)



3.19 Congenital dislocation of the hip

- Congenital dislocation of the hip is a condition in which the femoral head is either displaced or displaceable from the acetabulum of the pelvis. Many newborn infants have clinical findings such as the so-called 'clicky hip' that may be confused with congenital dislocation of the hip; there may be considerable variation in the terminology used to describe these findings.

- The International Classification of Diseases code for congenital dislocation of the hip is 754.3.

- The national rate of congenital dislocation of the hip in births increased from 18.6 per 10,000 births in 1985 to 25.4 per 10,000 births in 1990, and then declined to 18.1 per 10,000 births in 1994 (Table 3.37, Figure 3.37).

- Among 5,492 infants with congenital dislocation of the hip and known outcome, 0.4% were stillborn; 1.0% of liveborn infants died in the neonatal period.

- Associated major malformations were reported in 5.5% of the births with congenital dislocation of the hip and another 0.7% had a chromosomal abnormality.

- Because of differences in ascertainment and in the terms used to describe congenital dislocation of the hip, it is known to vary markedly in different regions. For births in 1990 to 1994, the reported rates for congenital dislocation of the hip were highest in Queensland and South Australia (43.2 and 32.1 per 10,000 births, respectively) and lowest in the Australian Capital Territory, Tasmania and Western Australia (0.4, 6.3 and 8.9 per 10,000 births, respectively) (Table 3.38, Figure 3.38). There was only 1 reported birth in the Australian Capital Territory.

Table 3.37: *Congenital dislocation of hip by outcome and type of malformation, Australia, 1985-1994*

Outcome	1985	1986	1987	1988	1989	1990	1991	1992	1993	1994	1985-94
						Number					
Live births	446	524	509	546	569	666	629	565	543	474	5,471
Stillbirths	3	1	1	3	1	3	1	6	3		22
Total births*	449	525	510	549	572	670	630	574	549	474	5,502
Induced abortions	-	-	-	-	1	-	-	-	-	-	1
Neonatal deaths	6	5	6	7	9	8	3	5	4	4	57
						Rate per 10,000 births					
Total births	18.6	21.4	20.8	22.2	22.7	25.4	24.5	21.8	21.1	18.1	21.7
						Number					
Isolated	418	494	477	512	534	616	602	545	511	455	5,164
Associated	26	27	27	35	36	47	27	26	31	18	300
Chromosomal	5	4	6	2	2	7	1	3	7	1	38
						Rate per 10,000 births					
Isolated	17.3	20.2	19.4	20.7	21.2	23.3	23.5	20.7	19.6	17.4	20.4
Associated	1.1	1.1	1.1	1.4	1.4	1.8	1.1	1.0	1.2	0.7	1.2
Chromosomal	0.2	0.2	0.2	0.1	0.1	0.3	0.0	0.1	0.3	0.0	0.1

** Total includes 'not stated'*

Table 3.38: *Congenital dislocation of hip, States and Territories, 1990-1994*

Outcome	NSW	Vic	Qld	WA	SA	Tas	ACT	NT	Australia
					Number				
Live births	512	882	997	111	317	16	1	41	2,877
Stillbirths	2	1	1	1	2	6	-	-	13
Total births*	517	883	1,001	112	320	22	1	41	2,897
Induced abortions	-	-	-	-	-	-	-	-	-
					Rate per 10,000 births				
Total births	11.7	26.9	43.2	8.9	32.1	6.3	0.4	23.1	22.2
					Number				
Isolated	458	857	971	81	306	21	1	34	2,729
Associated	51	24	26	28	12	1	-	7	149
Chromosomal	8	2	4	3	2	-	-	-	19
					Rate per 10,000 births				
Isolated	10.3	26.1	41.9	6.4	30.7	6.0	0.4	19.1	20.9
Associated	1.2	0.7	1.1	2.2	1.2	0.3	-	3.9	1.1
Chromosomal	0.2	0.1	0.2	0.2	0.2	-	-	-	0.1

** Total includes 'not stated'*

Figure 3.37: Congenital dislocation of hip, Australia, 1985-1994

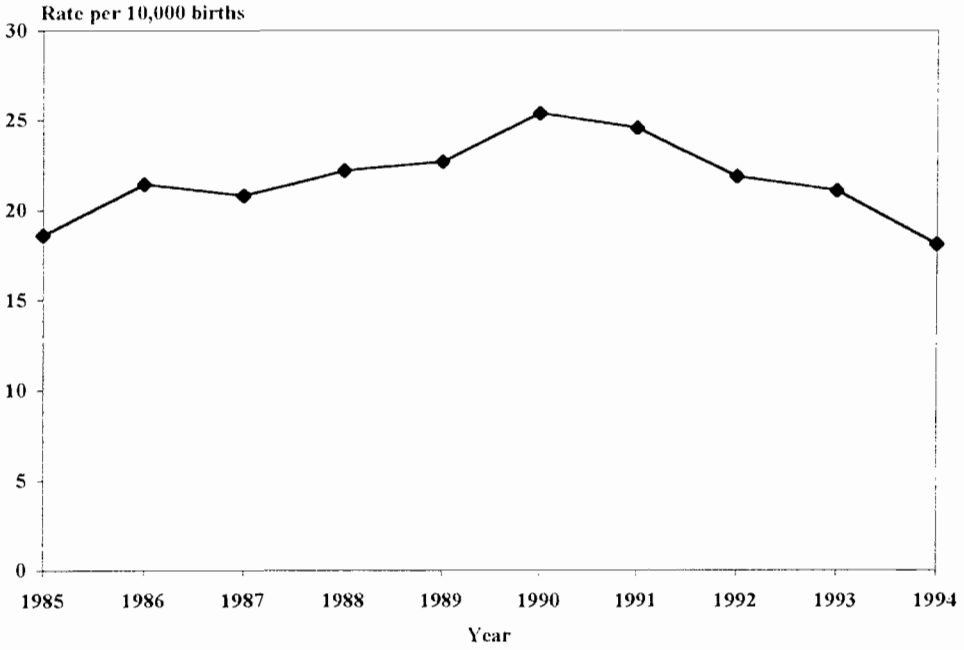
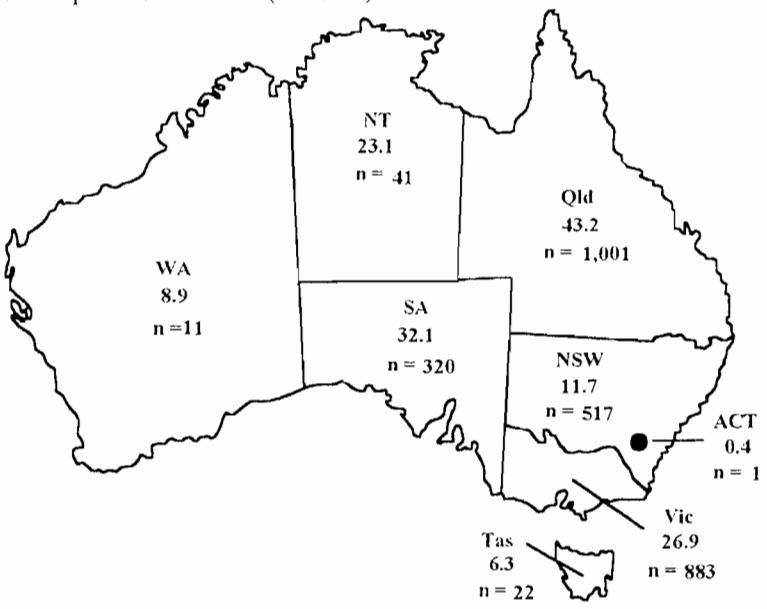


Figure 3.38: Congenital dislocation of hip, States and Territories, 1990-1994

Australia: 22.2 per 10,000 births (n=2,897)



3.20 Limb reduction defects

- Limb reduction defects are congenital malformations characterised by total or partial absence or severe hypoplasia of skeletal structures of the limbs. These are heterogeneous in type and are often analysed in the following groups: preaxial longitudinal, postaxial longitudinal, transverse, intercalary, multiple and unspecified.

- The International Classification of Diseases codes for limb reduction defects are 755.2-755.4. The NPSU coding of limb reduction defects was modified to include the groups specified above.

- From 1985 to 1994 the national rate of limb reduction defects in births varied between 5.2 per 10,000 births in 1989 and 3.9 per 10,000 births in 1985 and 1994 (Table 3.39, Figure 3.39).

- In the years 1985-1994, induced abortions were reported in 5.0% of all recorded notifications of limb reduction defects.

- Among 1,150 infants with limb reduction defects and known outcome, 13.2% were stillborn; 11.9% of liveborn infants died in the neonatal period.

- Associated major malformations were reported in 30.3% of the births with limb reduction defects and another 6.8% had a chromosomal abnormality.

- For births in 1990 to 1994, the reported rates for limb reduction defects were highest in South Australia (6.6 per 10,000 births) and lowest in the Australian Capital Territory and Tasmania (2.1 and 2.9 per 10,000 births, respectively) (Table 3.40, Figure 3.40).

Table 3.39: *Limb reduction defects by outcome and type of malformation, Australia, 1985-1994*

Outcome	1985	1986	1987	1988	1989	1990	1991	1992	1993	1994	1985-94
						Number					
Live births	84	100	83	105	115	99	113	114	99	86	998
Stillbirths	9	9	17	14	17	21	13	18	21	13	152
Total births*	93	109	100	120	132	120	128	133	120	101	1,156
Induced abortions	2	-	-	6	7	6	5	8	11	16	61
Neonatal deaths	12	17	20	8	15	13	8	7	8	11	119
						Rate per 10,000 births					
Total births	3.9	4.4	4.1	4.8	5.2	4.5	5.0	5.1	4.6	3.9	4.6
						Number					
Isolated	60	68	55	89	80	68	87	85	78	57	727
Associated	26	29	36	27	40	44	33	41	35	39	350
Chromosomal	7	12	9	4	12	8	8	7	7	5	79
						Rate per 10,000 births					
Isolated	2.5	2.8	2.2	3.6	3.2	2.6	3.4	3.2	3.0	2.2	2.9
Associated	1.1	1.2	1.5	1.1	1.6	1.7	1.3	1.6	1.3	1.5	1.4
Chromosomal	0.3	0.5	0.4	0.2	0.5	0.3	0.3	0.3	0.3	0.2	0.3

* *Total includes 'not stated'*

Table 3.40: *Limb reduction defects, States and Territories, 1990-1994*

Outcome	NSW	Vic	Qld	WA	SA	Tas	ACT	NT	Australia
					Number				
Live births	196	100	107	36	53	7	4	8	511
Stillbirths	23	31	10	7	11	3	1	-	86
Total births*	220	131	119	43	66	10	5	8	602
Induced abortions	9	10	-	11	12	-	4	-	46
					Rate per 10,000 births				
Total births	5.0	4.0	5.1	3.4	6.6	2.9	2.1	4.5	4.6
					Number				
Isolated	130	81	87	22	43	6	2	4	375
Associated	78	39	27	18	21	4	1	4	192
Chromosomal	12	11	5	3	2	-	2	-	35
					Rate per 10,000 births				
Isolated	2.9	2.5	3.8	1.7	4.3	1.7	0.8	2.3	2.9
Associated	1.8	1.2	1.2	1.4	2.1	1.2	0.4	2.3	1.5
Chromosomal	0.3	0.3	0.2	0.2	0.2	-	0.8	-	0.3

* *Total includes 'not stated'*

Figure 3.39: Limb reduction defects, Australia, 1985-1994

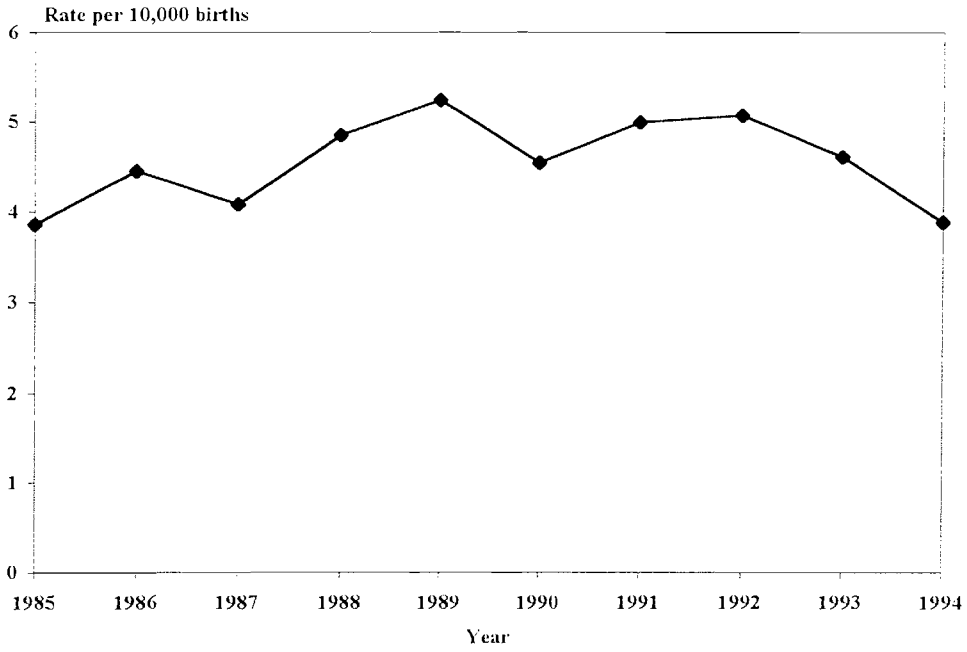
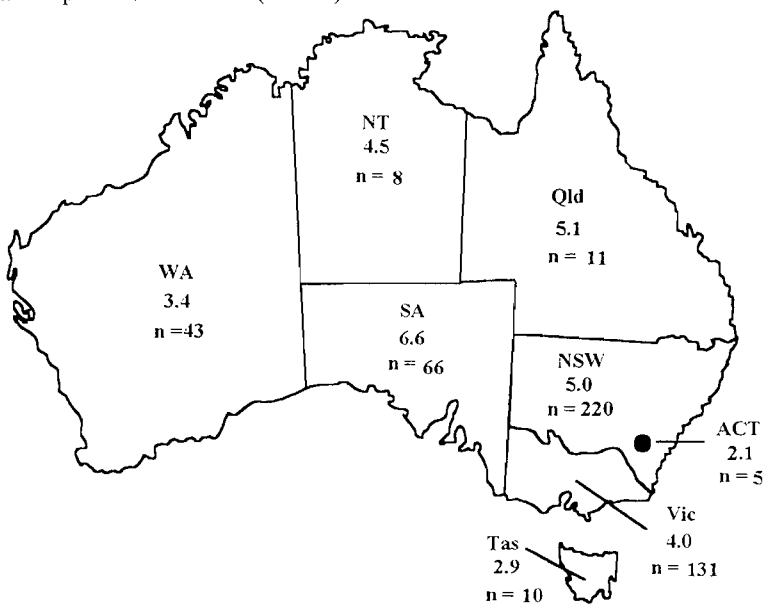


Figure 3.40: Limb reduction defects, States and Territories, 1990-1994

Australia: 4.6 per 10,000 births (n=602)



3.21 Diaphragmatic hernia

- Diaphragmatic hernia is a congenital malformation characterised by herniation into the thorax of abdominal contents through a defect of the diaphragm, but excluding eventration of the diaphragm.

- The International Classification of Diseases code for diaphragmatic hernia is 756.6.

- The national rate of diaphragmatic hernia in births varied between 2.1 (1988 and 1993) and 3.8 (1991) per 10,000 births, with an overall rate of 2.9 per 10,000 births during 1985 to 1994 (Table 3.41, Figure 3.41).

- In the years 1985-1994, induced abortions were reported in 5.1% of all recorded notifications of diaphragmatic hernia.

- Among 725 infants with diaphragmatic hernia and known outcome, 10.2% were stillborn; 51.5% of liveborn infants died in the neonatal period.

- Associated major malformations were reported in 25.3% of the births with diaphragmatic hernia and another 6.2% had a chromosomal abnormality.

- For births in 1990 to 1994, the reported rates for diaphragmatic hernia were highest in Queensland (3.7 per 10,000 births) and lowest in the Northern Territory (0.6 per 10,000 births) (Table 3.42, Figure 3.42). There was only 1 reported birth in the Northern Territory.

Table 3.41: *Diaphragmatic hernia by outcome and type of malformation, Australia, 1985-1994*

Outcome	1985	1986	1987	1988	1989	1990	1991	1992	1993	1994	1985-94
						Number					
Live births	69	67	69	49	65	77	83	70	48	54	651
Stillbirths	4	10	5	4	4	8	15	12	6	6	74
Total births*	73	77	74	53	69	85	98	82	54	61	726
Induced abortions	1	2	-	2	1	5	6	8	6	8	39
Neonatal deaths	46	38	50	27	37	38	42	27	11	19	335
						Rate per 10,000 births					
Total births	3.0	3.1	3.0	2.1	2.7	3.2	3.8	3.1	2.1	2.3	2.9
						Number					
Isolated	43	52	43	28	51	63	74	65	37	41	497
Associated	28	21	28	17	16	19	16	11	13	15	184
Chromosomal	2	4	3	8	2	3	8	6	4	5	45
						Rate per 10,000 births					
Isolated	1.8	2.1	1.8	1.1	2.0	2.4	2.9	2.5	1.4	1.6	2.0
Associated	1.2	0.9	1.1	0.7	0.6	0.7	0.6	0.4	0.5	0.6	0.7
Chromosomal	0.1	0.2	0.1	0.3	0.1	0.1	0.3	0.2	0.2	0.2	0.2

** Total includes 'not stated'*

Table 3.42: *Diaphragmatic hernia, States and Territories, 1990-1994*

Outcome	NSW	Vic	Qld	WA	SA	Tas	ACT	NT	Australia
					Number				
Live births	100	92	81	24	22	10	2	1	332
Stillbirths	8	20	4	6	6	1	2	-	47
Total births*	108	112	86	30	28	11	4	1	380
Induced abortions	3	12	-	5	13	-	-	-	33
					Rate per 10,000 births				
Total births	2.4	3.4	3.7	2.4	2.8	3.2	1.7	0.6	2.9
					Number				
Isolated	79	80	72	22	19	7	1	-	280
Associated	24	20	11	7	7	4	-	1	74
Chromosomal	5	12	3	1	2	-	3	-	26
					Rate per 10,000 births				
Isolated	1.8	2.4	3.1	1.7	1.9	2.0	0.4	-	2.1
Associated	0.5	0.6	0.5	0.6	0.7	1.2	-	0.6	0.6
Chromosomal	0.1	0.4	0.1	0.1	0.2	-	1.3	-	0.2

** Total includes 'not stated'*

Figure 3.41: Diaphragmatic hernia, Australia, 1985-1994

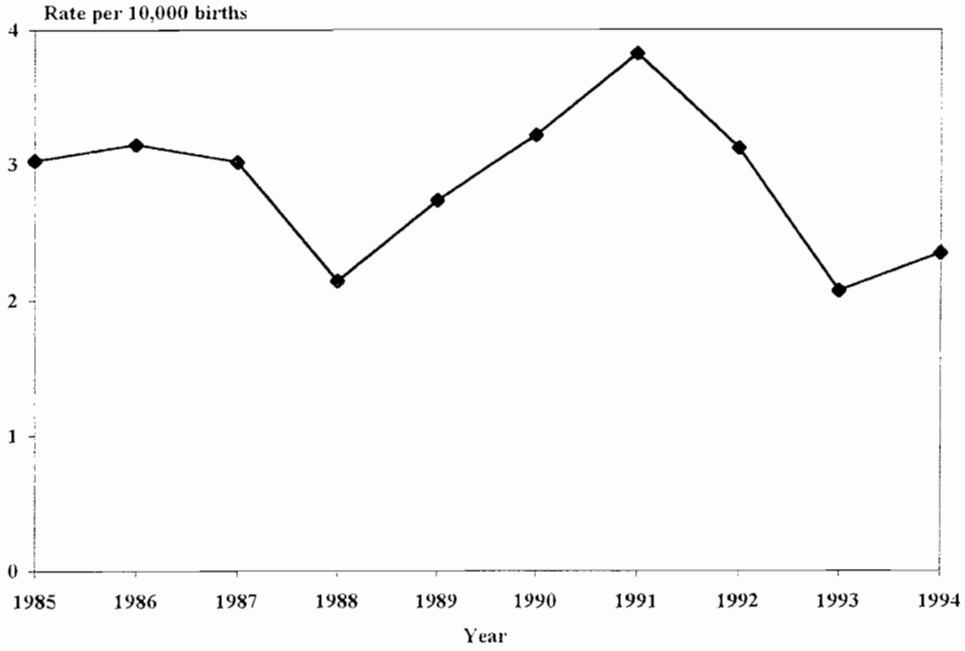
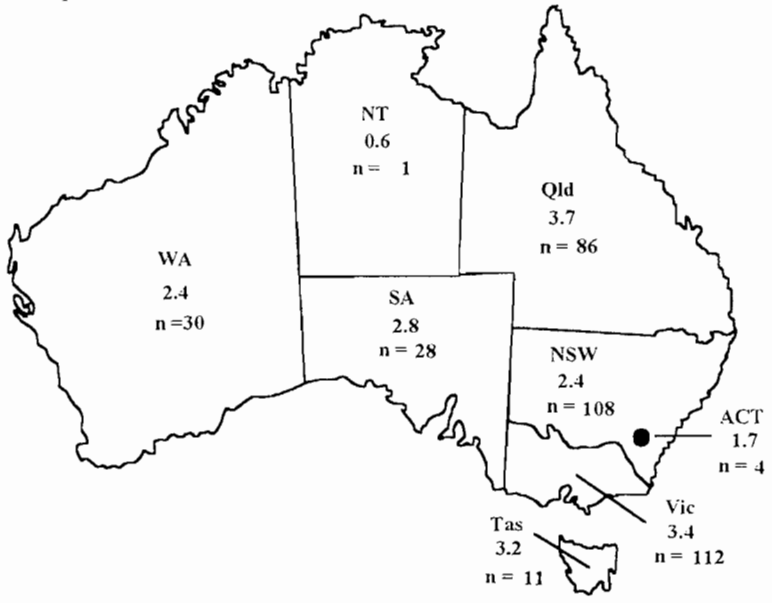


Figure 3.42: Diaphragmatic hernia, States and Territories, 1990-1994

Australia: 2.9 per 10,000 births (n=380)



3.22 Exomphalos

- Exomphalos is a congenital malformation characterised by herniation of the abdominal contents through the umbilical insertion and covered by a membrane which may or may not remain intact. Omphalocele is another term used to describe the same malformation.

- In the International Classification of Diseases, exomphalos is included under the code for anomalies of the abdominal wall (756.7). The British Paediatric Association code is 756.70, enabling distinction between exomphalos and other abdominal wall defects such as gastroschisis.

- The national rate of exomphalos in births has fallen by more than half from 2.7 per 10,000 births in 1986 to 1.1 per 10,000 births in 1994 (Table 3.43, Figure 3.43). During the same period, the number of induced abortions performed before 20 weeks' gestation for exomphalos increased.

- In the years 1985-1994, induced abortions were reported in 16.5% of all recorded notifications of exomphalos.

- Among 537 infants with exomphalos and known outcome, 30.7% were stillborn; 30.9% of liveborn infants died in the neonatal period.

- Associated major malformations were reported in 35.7% of the births with exomphalos and another 17.4% had a chromosomal abnormality.

- For births in 1990 to 1994, the reported rates of exomphalos were highest in South Australia and Western Australia (2.6 and 2.4 per 10,000 births, respectively) and lowest in the Northern Territory (0.6 per 10,000 births) (Table 3.44, Figure 3.44). There was only 1 reported birth in the Northern Territory.

Table 3.43: *Exomphalos by outcome and type of malformation, Australia, 1985-1994*

Outcome	1985	1986	1987	1988	1989	1990	1991	1992	1993	1994	1985-94
						Number					
Live births	48	51	32	33	39	48	36	32	33	20	372
Stillbirths	14	15	17	16	23	22	18	16	14	10	165
Total births*	62	66	49	49	62	71	56	48	47	30	540
Induced abortions	1	2	4	4	13	10	12	13	24	24	107
Neonatal deaths	21	23	11	11	13	11	10	3	9	3	115
						Rate per 10,000 births					
Total births	2.6	2.7	2.0	2.0	2.5	2.7	2.2	1.8	1.8	1.1	2.1
						Number					
Isolated	37	32	18	22	28	36	25	22	21	12	253
Associated	17	20	25	19	22	27	24	15	11	13	193
Chromosomal	8	14	6	8	12	8	7	11	15	5	94
						Rate per 10,000 births					
Isolated	1.5	1.3	0.7	0.9	1.1	1.4	1.0	0.8	0.8	0.5	1.0
Associated	0.7	0.8	1.0	0.8	0.9	1.0	0.9	0.6	0.4	0.5	0.8
Chromosomal	0.3	0.6	0.2	0.3	0.5	0.3	0.3	0.4	0.6	0.2	0.4

** Total includes 'not stated'*

Table 3.44: *Exomphalos, States and Territories, 1990-1994*

Outcome	NSW	Vic	Qld	WA	SA	Tas	ACT	NT	Australia
					Number				
Live births	52	41	27	28	15	5	1	-	169
Stillbirths	24	29	11	3	9	1	2	1	80
Total births*	77	70	38	31	26	6	3	1	252
Induced abortions	22	32	2	11	14	-	2	-	83
					Rate per 10,000 births				
Total births	1.7	2.1	1.6	2.4	2.6	1.7	1.3	0.6	1.9
					Number				
Isolated	32	33	19	15	11	3	3	-	116
Associated	33	23	10	12	12	-	-	-	90
Chromosomal	12	14	9	4	3	3	-	1	46
					Rate per 10,000 births				
Isolated	0.7	1.0	0.8	1.2	1.1	0.9	1.3	-	0.9
Associated	0.7	0.7	0.4	0.9	1.2	-	-	-	0.7
Chromosomal	0.3	0.4	0.4	0.3	0.3	0.9	-	0.6	0.4

** Total includes 'not stated'*

Figure 3.43: Exomphalos, Australia, 1985-1994

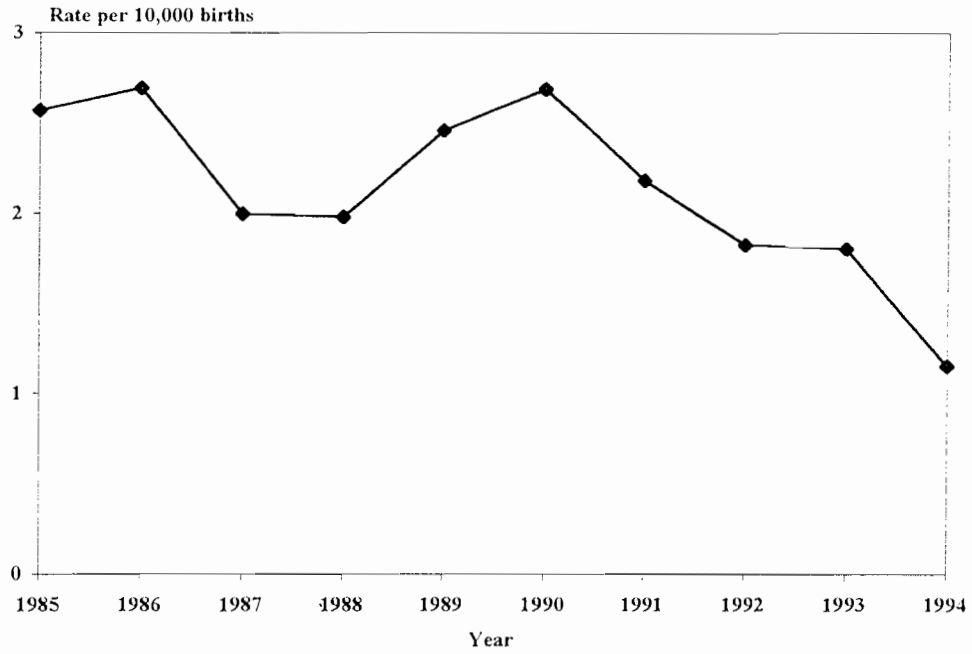
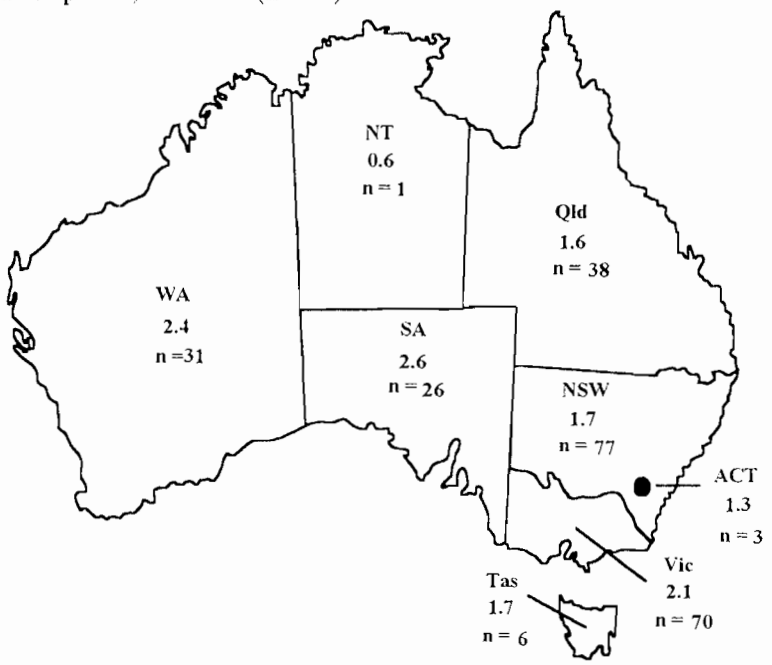


Figure 3.44: Exomphalos, States and Territories, 1990-1994

Australia: 1.9 per 10,000 births (n=252)



3.23 Gastroschisis

- Gastroschisis is a congenital malformation (para-umbilical hernia) characterised by visceral herniation through an abdominal wall defect lateral to an intact umbilical cord.

- In the International Classification of Diseases, gastroschisis is included under the code for anomalies of the abdominal wall (756.7). The British Paediatric Association code is 756.71, enabling distinction between gastroschisis and other abdominal wall defects such as exomphalos.

- The national rate of gastroschisis in births increased during the period from 1985 to 1994 and varied between 0.8 per 10,000 births in 1985 and 2.0 per 10,000 births in 1993 (Table 3.45, Figure 3.45). There were relatively few induced abortions for gastroschisis during this period.

- In the years 1985-1994, induced abortions were reported in 5.8% of all recorded notifications of gastroschisis.

- Among 343 infants with gastroschisis and known outcome, 11.4% were stillborn; 6.6% of liveborn infants died in the neonatal period.

- Associated major malformations were reported in 14.8% of the births with gastroschisis and 1 infant had a chromosomal abnormality.

- For births in 1990 to 1994, the reported rates for gastroschisis were highest in Western Australia and Queensland (2.1 and 1.9 per 10,000 births, respectively) and lowest in the Northern Territory (0.6 per 10,000 births) (Table 3.46, Figure 3.46). There was only 1 reported birth in the Northern Territory.

Table 3.45: *Gastroschisis by outcome and type of malformation, Australia, 1985-1994*

Outcome	1985	1986	1987	1988	1989	1990	1991	1992	1993	1994	1985-94
						Number					
Live births	18	16	30	32	27	21	34	41	47	38	304
Stillbirths	2	6	5	3	6	1	3	5	6	2	39
Total births*	20	22	35	35	33	23	37	46	53	40	344
Induced abortions	-	-	1	2	-	3	2	3	3	7	21
Neonatal deaths	2	1	6	5	1	-	1	3	1	-	20
						Rate per 10,000 births					
Total births	0.8	0.9	1.4	1.4	1.3	0.9	1.4	1.8	2.0	1.5	1.4
						Number					
Isolated	17	17	22	28	27	21	31	37	52	40	292
Associated	3	5	13	7	5	2	6	9	1	-	51
Chromosomal	-	-	-	-	1	-	-	-	-	-	1
						Rate per 10,000 births					
Isolated	0.7	0.7	0.9	1.1	1.1	0.8	1.2	1.4	2.0	1.5	1.2
Associated	0.1	0.2	0.5	0.3	0.2	0.1	0.2	0.3	0.0	-	0.2
Chromosomal	-	-	-	-	0.0	-	-	-	-	-	0.0

* *Total includes 'not stated'*

Table 3.46: *Gastroschisis, States and Territories, 1990-1994*

Outcome	NSW	Vic	Qld	WA	SA	Tas	ACT	NT	Australia
					Number				
Live births	59	36	42	23	15	3	2	1	181
Stillbirths	3	5	1	4	1	2	1	-	17
Total births*	63	41	43	27	16	5	3	1	199
Induced abortions	4	6	-	1	3	1	3	-	18
					Rate per 10,000 births				
Total births	1.4	1.2	1.9	2.1	1.6	1.4	1.3	0.6	1.5
					Number				
Isolated	56	33	43	24	16	5	3	1	181
Associated	7	8	-	3	-	-	-	-	18
Chromosomal	-	-	-	-	-	-	-	-	-
					Rate per 10,000 births				
Isolated	1.3	1.0	1.9	1.9	1.6	1.4	1.3	0.6	1.4
Associated	0.2	0.2	-	0.2	-	-	-	-	0.1
Chromosomal	-	-	-	-	-	-	-	-	-

* *Total includes 'not stated'*

Figure 3.45: Gastroschisis, Australia, 1985-1994

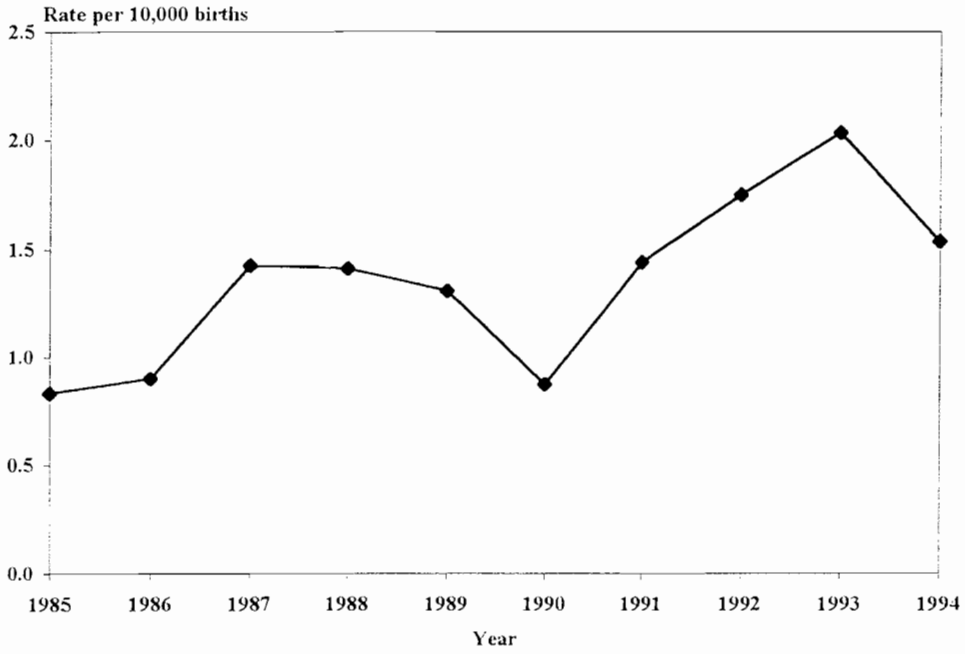
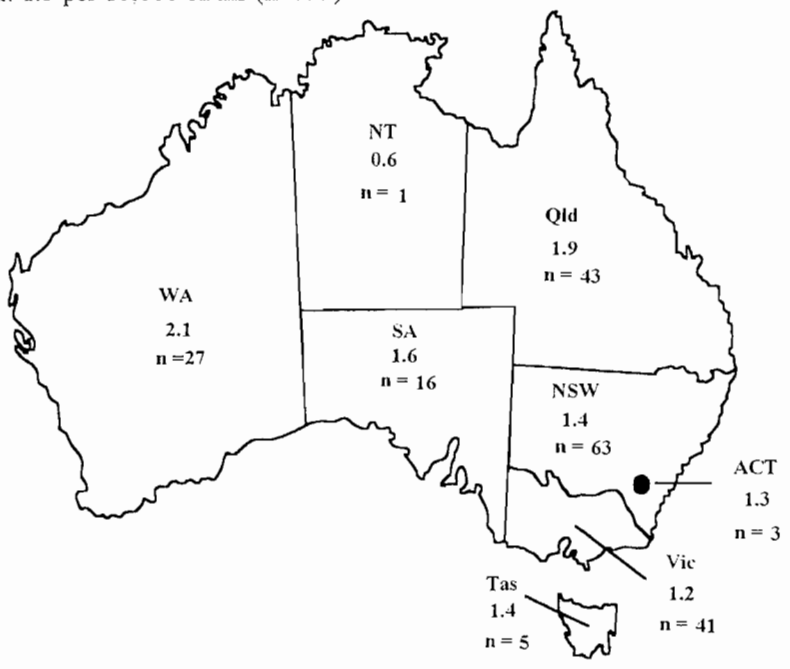


Figure 3.46: Gastroschisis, States and Territories, 1990-1994

Australia: 1.5 per 10,000 births (n=199)



3.24 Trisomy 21 (Down syndrome)

- Trisomy 21 (Down syndrome) is characterised by a specific pattern of abnormalities including hypotonia, flat facies, slanted palpebral fissures, small ears, intellectual disability and variable occurrence of other minor and major congenital malformations. On chromosomal analysis, there is an additional chromosome 21 or part of its long arm.

- The International Classification of Diseases code for Down syndrome is 758.0. The 5-digit British Paediatric Association Classification enables separate codes for the different types of chromosomal abnormality (trisomy 21, translocation, mosaic).

- The national rate of Down syndrome in births was relatively constant at around 12.8 per 10,000 births during the period 1985 to 1994, ranging between a high of 14.0 per 10,000 births in 1993 and a low of 11.9 per 10,000 births in 1987 (Table 3.47, Figure 3.47). The reported number of induced abortions performed after prenatal diagnosis of trisomy 21 by amniocentesis or chorionic villus sampling increased substantially during this period, reaching the highest number of 128 in 1994.

- In the years 1985-1994, induced abortions were reported in 18.3% of all recorded notifications of Down syndrome.

- Among 3,148 infants with trisomy 21 and known outcome, 8.8% were stillborn; 4.2% of liveborn infants died in the neonatal period.

- For births in 1990 to 1994, the reported rates for Down syndrome were highest in the Australian Capital Territory, New South Wales and Victoria (13.9, 13.6 and 13.6 per 10,000 births, respectively) and lowest in the Northern Territory (9.6 per 10,000 births) (Table 3.48, Figure 3. 48).

Table 3.47: *Trisomy 21 by outcome and type of malformation, Australia, 1985-1994*

Outcome	1985	1986	1987	1988	1989	1990	1991	1992	1993	1994	1985-94
						Number					
Live births	270	302	265	282	306	312	305	278	291	261	2,872
Stillbirths	23	27	24	21	19	29	26	41	25	41	276
Total births*	293	330	292	305	327	345	336	320	366	320	3,234
Induced abortions	26	35	46	55	37	71	94	113	118	128	723
Neonatal deaths	14	15	16	8	24	8	12	5	7	12	121
						Rate per 10,000 births					
Total births	12.2	13.5	11.9	12.3	13.0	13.1	13.1	12.2	14.0	12.3	12.8
						Number					
Isolated	-	-	-	-	-	-	-	-	-	-	-
Associated	-	-	-	-	-	-	-	-	-	-	-
Chromosomal	293	330	292	305	327	345	336	320	366	320	3,234
						Rate per 10,000 births					
Isolated	-	-	-	-	-	-	-	-	-	-	-
Associated	-	-	-	-	-	-	-	-	-	-	-
Chromosomal	12.2	13.5	11.9	12.3	13.0	13.1	13.1	12.2	14.0	12.3	12.8

** Total includes 'not stated'*

Table 3.48: *Trisomy 21, States and Territories, 1990-1994*

Outcome	NSW	Vic	Qld	WA	SA	Tas	ACT	NT	Australia
					Number				
Live births	493	398	279	129	71	35	27	15	1,447
Stillbirths	39	43	24	13	29	7	5	2	162
Total births*	600	446	303	142	104	42	33	17	1,687
Induced abortions	203	153	47	57	43	9	10	2	524
					Rate per 10,000 births				
Total births	13.6	13.6	13.1	11.2	10.4	12.1	13.9	9.6	12.9
					Number				
Isolated	-	-	-	-	-	-	-	-	-
Associated	-	-	-	-	-	-	-	-	-
Chromosomal	600	446	303	142	104	42	33	17	1,687
					Rate per 10,000 births				
Isolated	-	-	-	-	-	-	-	-	-
Associated	-	-	-	-	-	-	-	-	-
Chromosomal	13.6	13.6	13.1	11.2	10.4	12.1	13.9	9.6	12.9

** Total includes 'not stated'*

Figure 3.47: Trisomy 21, Australia, 1985-1994

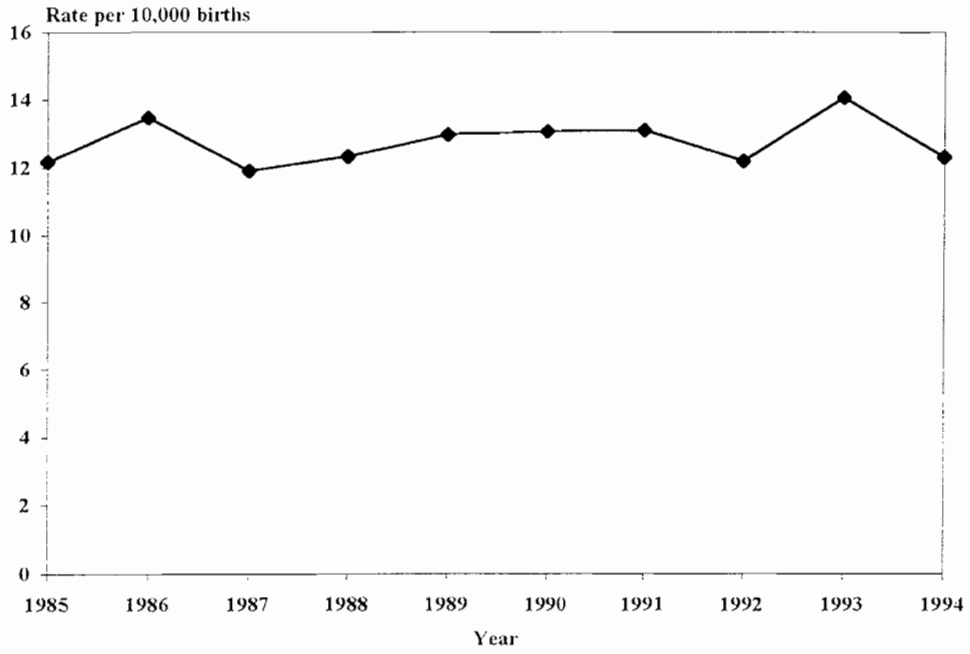
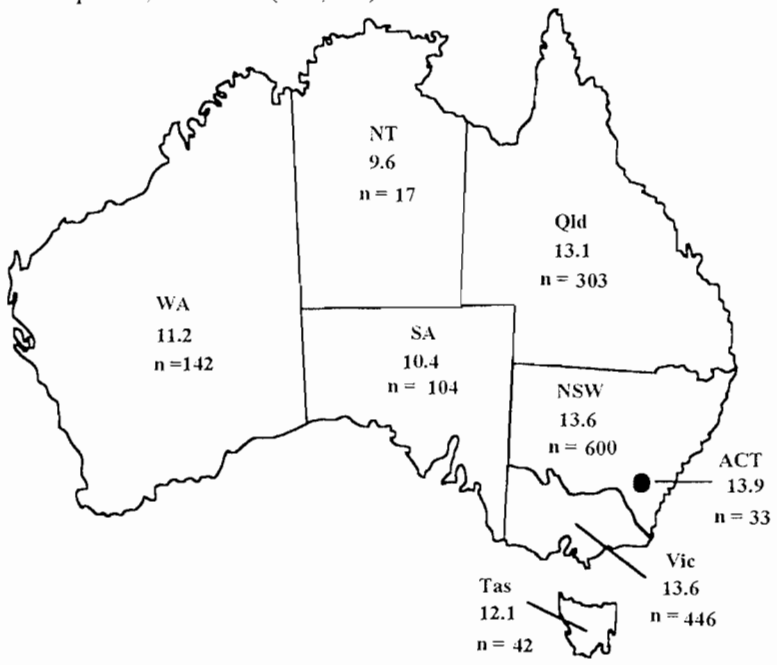


Figure 3.48: Trisomy 21, States and Territories, 1990-1994

Australia: 12.9 per 10,000 births (n=1,687)



3.25 Trisomy 18 (Edwards syndrome)

- Trisomy 18 (Edwards syndrome) is characterised by a specific pattern of abnormalities including clenched hand and overlapping fingers, abnormal dermal ridge pattern of the fingertips, developmental disability and variable occurrence of other minor and major congenital malformations. On chromosomal analysis, there is an additional chromosome 18 or part of this chromosome.

- The International Classification of Diseases code for Edwards syndrome is 758.2. The 5-digit British Paediatric Association Classification enables separate codes for the different types of chromosomal abnormality (trisomy 18, translocation, mosaic).

- The national rate of Edwards syndrome in births was relatively constant at around 2.2 per 10,000 births during the period 1985 to 1994, ranging between a high of 2.8 per 10,000 births in 1991 and a low of 1.8 per 10,000 births in 1986 (Table 3.49, Figure 3.49). The reported number of induced abortions performed after prenatal diagnosis of trisomy 18 by amniocentesis or chorionic villus sampling increased substantially during this period, reaching the highest number of 48 in 1994.

- In the years 1985-1994, induced abortions were reported in 29.6% of all recorded notifications of Edwards syndrome.

- Among 541 infants with trisomy 18 and known outcome, 33.1% were stillborn; 64.1% of liveborn infants died in the neonatal period.

- For births in 1990 to 1994, the reported rates for Edwards syndrome were highest in the Northern Territory, New South Wales and Victoria (2.8, 2.7 and 2.6 per 10,000 births, respectively) and lowest in South Australia (1.6 per 10,000 births) (Table 3.50, Figure 3.50).

Table 3.49: *Trisomy 18 by outcome and type of malformation, Australia, 1985-1994*

Outcome	1985	1986	1987	1988	1989	1990	1991	1992	1993	1994	1985-94
						Number					
Live births	47	31	35	41	30	37	50	37	30	24	362
Stillbirths	12	11	12	12	18	21	21	21	25	26	179
Total births*	59	43	47	54	49	61	72	58	71	54	568
Induced abortions	13	11	11	12	21	20	29	40	34	48	239
Neonatal deaths	33	23	28	26	22	22	33	17	15	13	232
						Rate per 10,000 births					
Total births	2.4	1.8	1.9	2.2	1.9	2.3	2.8	2.2	2.7	2.1	2.2
						Number					
Isolated	-	-	-	-	-	-	-	-	-	-	-
Associated	-	-	-	-	-	-	-	-	-	-	-
Chromosomal	59	43	47	54	49	61	72	58	71	54	568
						Rate per 10,000 births					
Isolated	-	-	-	-	-	-	-	-	-	-	-
Associated	-	-	-	-	-	-	-	-	-	-	-
Chromosomal	2.4	1.8	1.9	2.2	1.9	2.3	2.8	2.2	2.7	2.1	2.2

* *Total includes 'not stated'*

Table 3.50: *Trisomy 18, States and Territories, 1990-1994*

Outcome	NSW	Vic	Qld	WA	SA	Tas	ACT	NT	Australia
					Number				
Live births	65	50	29	14	6	6	4	4	178
Stillbirths	35	34	22	9	10	1	2	1	114
Total births*	121	86	51	24	16	7	6	5	316
Induced abortions	57	65	11	19	11	1	7	-	171
					Rate per 10,000 births				
Total births	2.7	2.6	2.2	1.9	1.6	2.0	2.5	2.8	2.4
					Number				
Isolated	-	-	-	-	-	-	-	-	-
Associated	-	-	-	-	-	-	-	-	-
Chromosomal	121	86	51	24	16	7	6	5	316
					Rate per 10,000 births				
Isolated	-	-	-	-	-	-	-	-	-
Associated	-	-	-	-	-	-	-	-	-
Chromosomal	2.7	2.6	2.2	1.9	1.6	2.0	2.5	2.8	2.4

* *Total includes 'not stated'*

Figure 3.49: Trisomy 18, Australia, 1985-1994

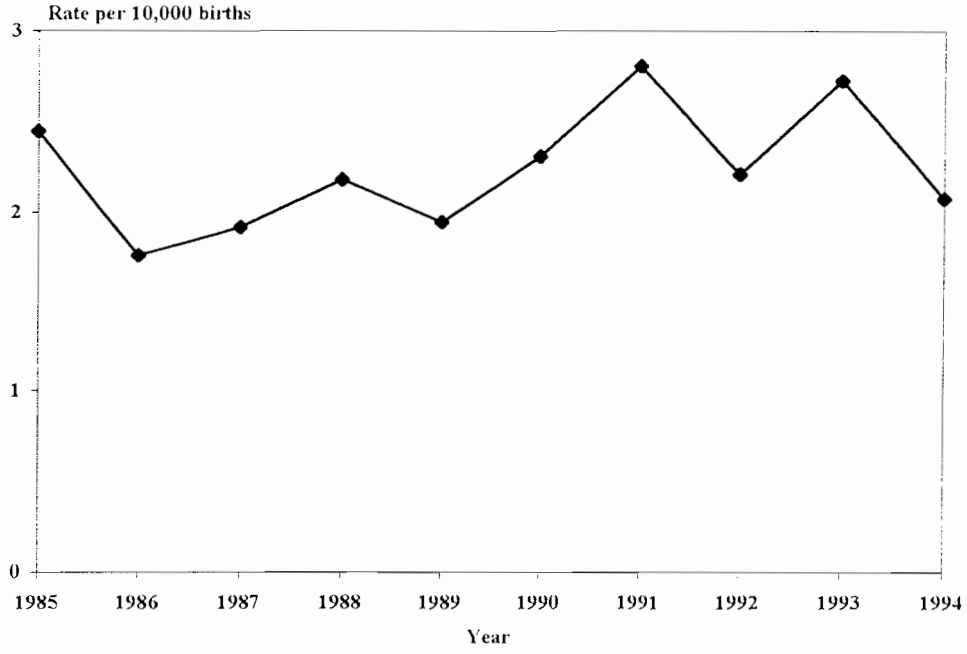
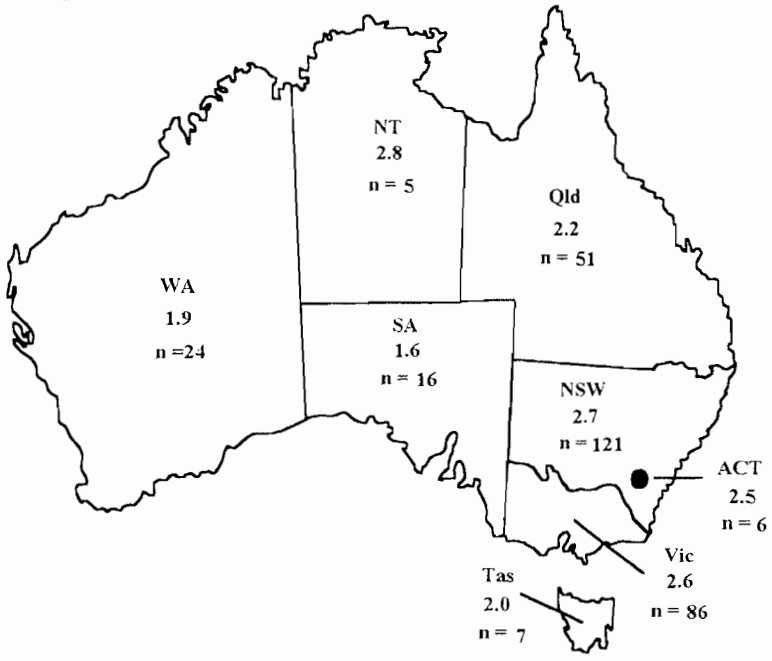


Figure 3.50: Trisomy 18, States and Territories, 1990-1994

Australia: 2.4 per 10,000 births (n=316)



4 Perinatal, infant and childhood deaths due to congenital malformations

Congenital malformations are an important cause of perinatal and infant deaths in Australia. Although the causes of most malformations remain unknown, their occurrence at birth is increasingly being influenced by prenatal diagnosis and possible termination of pregnancy if lethal or severe malformations are detected. Many induced abortions of malformed fetuses occur at gestational ages below the usual limits that define perinatal deaths. Without effective notification of induced abortions, it is likely that the public health significance of congenital malformations and their contribution to fetal and early childhood death will be underestimated.

National data on perinatal deaths and infant deaths from the Australian Bureau of Statistics (ABS) were analysed for the period 1973 to 1994, and 1980 to 1994, respectively. This analysis determined the overall trends in death rates for specific types of malformations, the trends in the proportion of perinatal deaths due to malformations for various gestational age and birthweight groups, and the relative proportion of deaths due to malformations in each age group.

4.1 Data and methods

There are differing legal and statistical definitions in Australia for registering and reporting perinatal deaths. For legal purposes, all fetal and neonatal deaths of at least 20 weeks' gestation or at least 400g birthweight are registered. The Australian Bureau of Statistics (ABS) publishes annual data on perinatal deaths based upon recommendations of the World Health Organization (WHO) for reporting national perinatal statistics. Fetal deaths are included if the birthweight is at least 500 grams or, when birthweight is not available, if the gestational age is at least 22 weeks, and there is no evidence of life after birth. Neonatal deaths are liveborn infants dying within 28 days of birth. Perinatal deaths include fetal deaths and neonatal deaths.

Fetal, neonatal and perinatal death numbers and rates in this section of the report are based on these ABS criteria. Denominators used in the calculation of death rates are annual births (live births for neonatal deaths and total births for fetal and perinatal death rates). Rates are expressed per 10,000 births in this report to maintain consistency with the malformation rates. Annual data are based on the year of registration.

National data on infant and childhood deaths from the ABS have also been analysed. An infant death is defined as the death of a live born child occurring within one year of birth. Childhood deaths are deaths in children aged between 1 and 14 years. Denominators used in the calculation of death rates are annual live births for infant deaths and resident population in the 1 to 14 year old age group for childhood deaths. Rates are expressed per 10,000 live births or 10,000 resident population.

Causes of death due to congenital malformations are classified according to the Ninth(1975) Revision of the World Health Organisation's International Classification of Diseases (ICD-9).

Trend lines have been fitted using a second order polynomial regression model to approximate a line of best fit (Kleinbaum, Kupper, Muller 1988).

4.2 Results

The overall perinatal death rate from congenital malformations declined from 35.9 per 10,000 births in 1973 to the lowest rate of 17.5 in 1994 (Figure 4.1). There was a marked fall in the death rate due to anencephalus, and lesser falls for the death rates due to spina bifida, hydrocephalus, and congenital heart defects. The perinatal death rates due to chromosomal abnormalities showed an increase in the same period, from 2.2 per 10,000 births in 1973 to 3.9 per 10,000 in 1994.

The proportion of perinatal deaths due to malformations showed an initially increasing trend after 1973, rising from 17% in that year to a peak of 26% in 1983, then fluctuating until 1994, when the proportion was 22% (Figure 4.2). A similar trend was evident for neonatal deaths, but the proportion of fetal deaths due to congenital malformations showed little change.

There was a gradual rise in the proportion of perinatal deaths that had relatively short gestational ages (Figure 4.3) or relatively low birthweights (Figure 4.4). These trends probably reflected an increasing number of terminations of pregnancy that followed prenatal diagnosis of fetal malformations.

Infant deaths from congenital malformations declined from 28.8 per 10,000 live births in 1980 to 17.6 10,000 in 1994 (Figure 4.5). This decline was more pronounced for malformations of the central nervous system than for congenital heart defects or other types of malformations. The proportion of infant deaths that were due to malformations also fluctuated during this period, varying from 26.8% in 1980 to 30.0% in 1994 with the highest proportion (30.6%) in 1983 and lowest proportion (24.6%) in 1989 (Figure 4.6).

The death rates of children aged 1-14 years from congenital malformations declined during the same period, from 3.6 per 10,000 children in 1980 to 2.2 per 10,000 in 1994 (Figure 4.5). The proportion of childhood deaths due to malformations rose slightly from 8.5% in 1980 to 10.2 % in 1994.

Figure 4.1: Trends in perinatal death rates for specific types of congenital malformations, Australia, 1973-1994

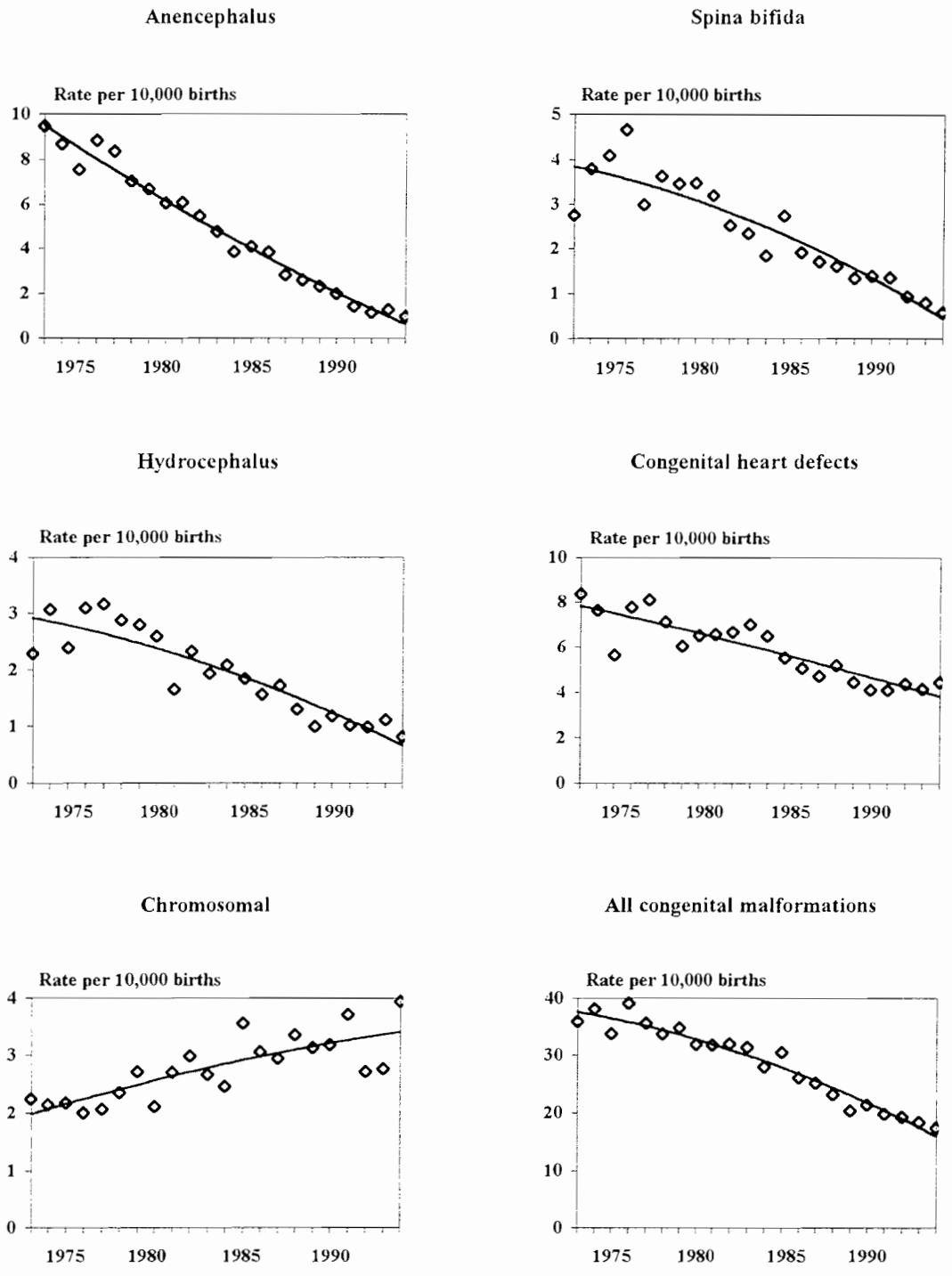


Figure 4.2: Proportion of fetal, neonatal and perinatal deaths due to congenital malformations, Australia, 1973-1994

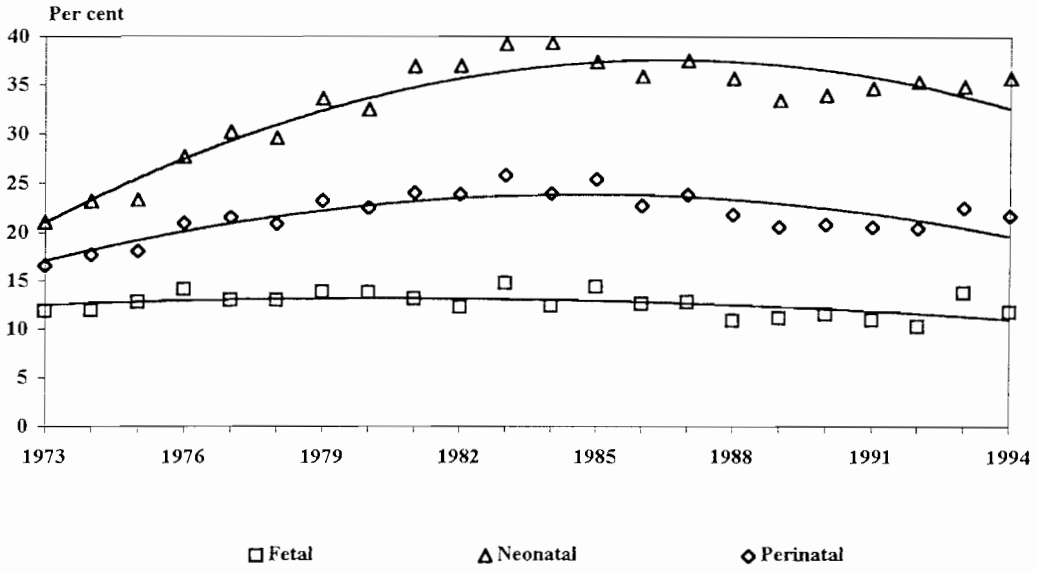


Figure 4.3: Proportion of perinatal deaths due to congenital malformations in selected gestational age groups, Australia, 1973-1994

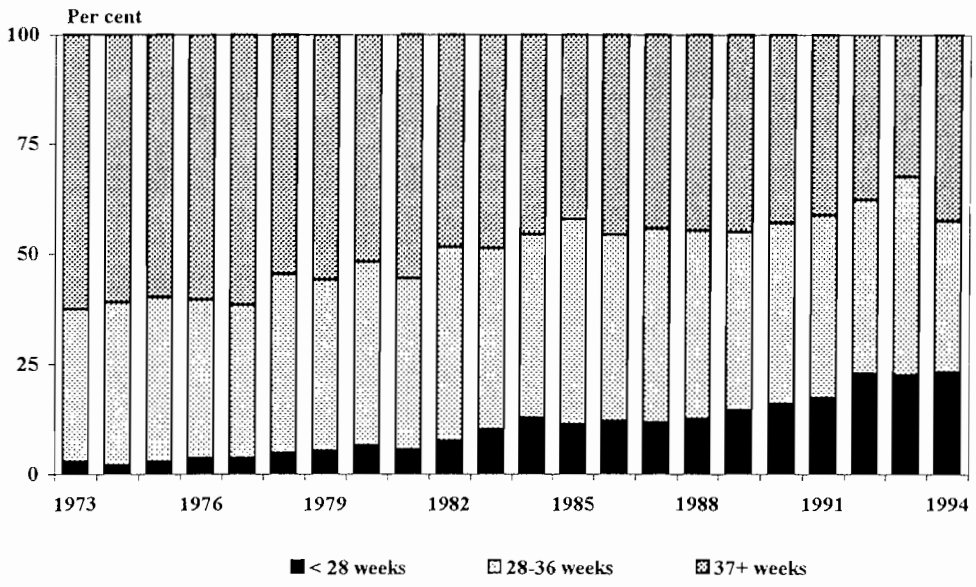


Figure 4.4: Proportion of perinatal deaths due to congenital malformations in selected birthweight groups, Australia, 1973-1994

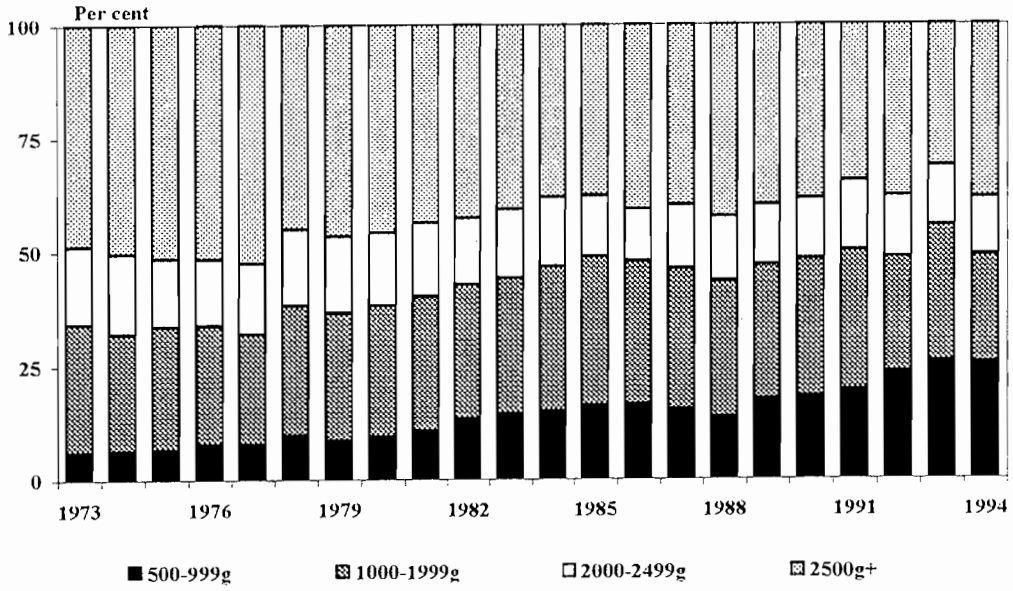
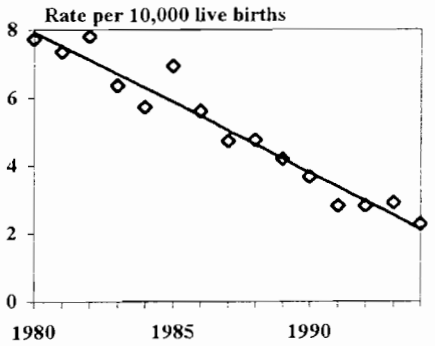
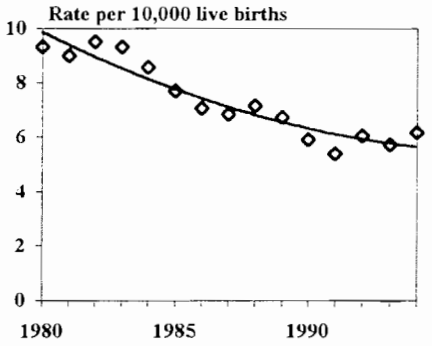


Figure 4.5: Trends in infant and childhood death rates for specific types of congenital malformations, Australia, 1980-1994

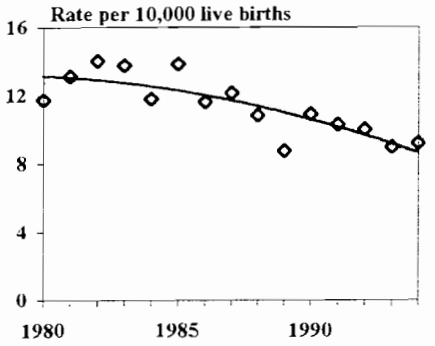
Central nervous system



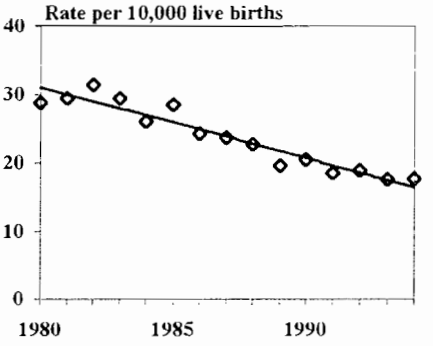
Congenital heart defects



Other systems



All congenital malformations



All childhood deaths (1-14 years)

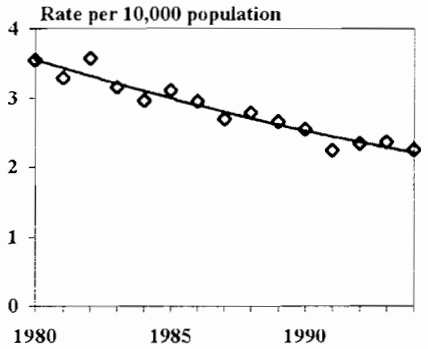
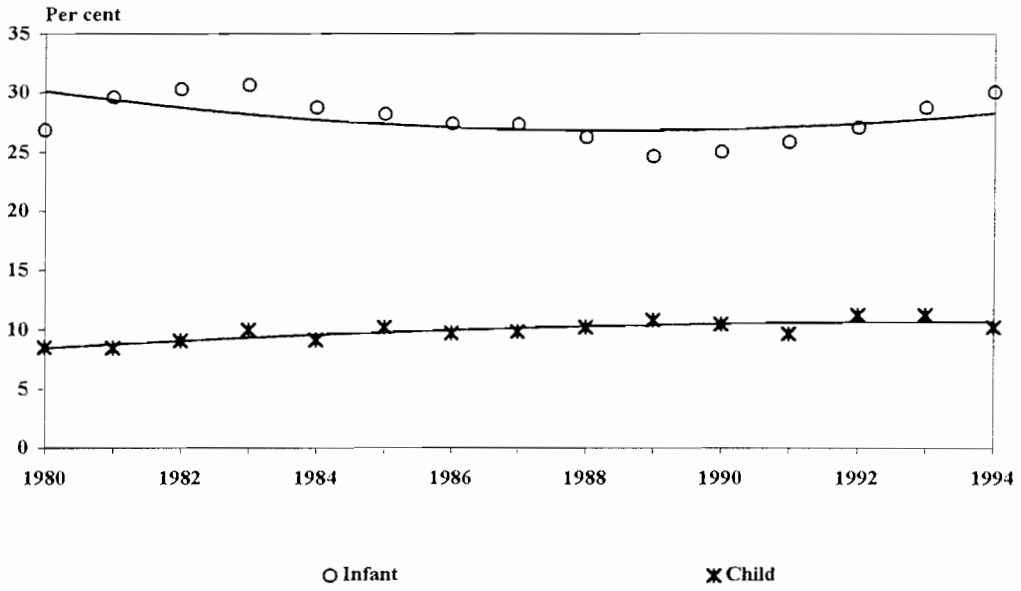


Figure 4.6: Proportion of infant and childhood deaths due to congenital malformations, Australia, 1980-1994



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Appendix 1 Minor congenital malformations

Unless an infant or fetus has other major congenital malformations, the following minor malformations and other conditions are not notified to the AIHW National Perinatal Statistics Unit.

Skin

- Skin cysts
- Noncavernous single small hemangioma (less than 10cm diameter)
- Benign skin neoplasms
- Nevus flammeus
- Birth mark
- Mongolian spots
- Cutis marmorata
- Café au lait spots
- Scalp defects, cutis aplasia
- Lanugo excessive or persistent
- Accessory nipple
- Pilonidal or sacral dimple

Skull

- Brachycephaly, dolichocephaly, plagiocephaly
- Craniotabes
- Large, small or absent fontanelles
- Macrocephaly
- Head asymmetry

Face

- Facial palsy
- Facial asymmetry
- Micrognathia
- Flat or wide nasal bridge, upturned nose, or other minor nose malformation
- Deviation of nasal septum

Eyes

- Esotropia, exotropia, strabismus
- Nystagmus
- Blue sclera
- Brushfield spots
- Epicanthal folds
- Eye slant (upward or downward)
- Narrow palpebral fissures
- Nasolacrimal duct obstruction/Dacryostenosis

Ears

- Ear tags
- Bat, cauliflower, elfin, lop, pointed, posteriorly rotated, or low-set ears
- Darwin's tubercle
- Preauricular sinus, cyst or pit
- Macrotia

Mouth, tongue and palate

- Tongue-tie
- Tongue cyst
- Ranula
- Cleft gum
- Macroglossia
- Microglossia
- Natal teeth
- Big, wide or small lips
- High-arched palate
- Bifid uvula

Neck

- Branchial cleft or sinus
- Redundant neck skin folds
- Webbing of neck
- Short neck

Cardiovascular system

- Patent ductus arteriosus or foramen ovale (gestational age <37 weeks or birthweight <2500g)
- Mild, trivial, or physiologic valvular regurgitation
- Cardiomegaly
- Dextroposition of heart
- Heart block
- Persistent fetal circulation
- Single umbilical artery

Gastrointestinal system

- Hepatomegaly
- Splenomegaly
- Meckel's diverticulum
- Anal tags
- Anal or rectal fissures
- Inguinal hernia in males
- Inguinal hernia in females (birthweight <2500g)
- Umbilical hernia (skin covered)

Urogenital system

- Imperforate hymen
- Prominent clitoris
- Fusion of vulva
- Vaginal or hymenal tags
- Cyst of vagina, vulva, canal of Nuck, or ovary
- Hydrocele
- Undescended testis (gestational age <37 weeks, birthweight <2500g)
- Small penis
- Chordee
- Patent urachus or urachal cyst
- Ectopic kidney

Limbs

- Skin tags on hands and feet
- Partial syndactyly of toes, webbing of toes
- Brachydactyly, unspecified
- Clinodactyly
- Camptodactyly
- Flexion deformities of digits
- Long fingers and toes
- Nail hypoplasia
- Enlarged or hypertrophic nails
- Widely spaced first and second toes
- Overlapping toes
- Tibial torsion or bowing
- Genu valgum, varum or recurvatum
- Dislocation of subluxation of knee
- Hallux valgus
- Hallux varus
- Talipes calcaneovalgus, equinovarus
- Cervical rib, other extra ribs
- Rocker-bottom feet
- Simian or Sydney lines, abnormal palmar creases
- Hip subluxation, clicky hips

Respiratory system

- Hypoplastic lungs (gestational age <37 weeks)
- Laryngeal stridor
- Laryngomalacia

Other conditions

- Balanced autosomal translocations
- Birth injuries
- Cephalhaematoma
- Cystic fibrosis
- Enzyme deficiencies
- Hydrops fetalis
- Meconium ileus
- Metabolic disorders
- Pyloric stenosis
- Sternomastoid tumour
- Torticollis
- Volvulus

Appendix 2 Data items in national monitoring system on congenital malformations

Demographic data:	State/Territory of birth Reference number State/Territory record number
Maternal data:	Local Government Area of residence Date of birth/Age Marital status Previous pregnancies and outcome Country of birth Aboriginality Accommodation status in hospital Date of last menstrual period
Infant/fetus data:	Hospital of birth (or termination of pregnancy) Sources of notification Date of birth (or termination of pregnancy) Sex Plurality and birth order Birthweight Gestational age Method of prenatal diagnosis (for aborted fetuses) Outcome Date of death (if applicable) Autopsy performed British Paediatric Association codes for congenital malformations Malformation type (eg isolated, multiple, syndrome) Sources of diagnosis Comment (optional), including cytogenetic diagnosis Follow-up information requested

Appendix 3 Definitions

Birthweight: The first weight of the baby (stillborn or liveborn) obtained after birth (usually measured to the nearest five grams and obtained within one hour of birth).

Congenital malformations: Structural or anatomical abnormalities that are present at birth, usually resulting from abnormal development in the first trimester of pregnancy.

Gestational age: The duration of pregnancy in completed weeks calculated from the date of a woman's last menstrual period and her infant's date of birth, or derived from clinical assessment during pregnancy or from examination of the infant after birth.

Induced abortion: Termination of pregnancy by medical or mechanical means before 20 weeks' gestation.

Infant death: Death of a liveborn 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 malformation is present in the infant or fetus.

Live birth: Live birth is 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 liveborn (WHO definition).

Low birthweight: Birthweight of less than 2500g.

Major congenital malformations: Congenital malformations that are either lethal or significantly affect the individual's function or appearance.

Maternal age: Mother's age at her child's birth.

Multiple malformations: More than one independent major congenital malformation is present in the infant or fetus.

Neonatal death: A death of a liveborn infant within 28 days of birth. Neonatal death rates are expressed per 10,000 live births in this report.

Perinatal death: Stillbirth (fetal death) or neonatal death. Perinatal death rates are expressed per 10,000 total births in this report.

Plurality: The number of births resulting from a pregnancy.

Preterm birth: Birth before 37 completed weeks of gestation.

Stillbirth (fetal death): Stillbirth is a fetal 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. Fetal death rates are expressed per 10,000 total births in this report.