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Congenital heart disease in Australia

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Australian Government

**Australian Institute of
Health and Welfare**

Congenital heart disease in Australia

Australian Institute of Health and Welfare
Canberra

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Australian Institute of Health and Welfare

Board Chair
Mrs Louise Markus

Chief Executive Officer
Mr Barry Sandison

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

Australian Institute of Health and Welfare

GPO Box 570

Canberra ACT 2601

Tel: (02) 6244 1000

Email: info@aihw.gov.au

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Summary

Congenital heart disease is a general term for any defect of the heart, heart valves or central blood vessels that is present at birth. Most congenital heart disease is multifactorial and arises through combinations of genetic and environmental factors. People with complex and severe congenital heart disease require specialist treatment throughout their life. This report presents a statistical overview of the number of people with congenital heart disease in Australia, on hospital treatment and care, and on mortality.

Globally, around 9 in every 1,000 babies born are affected by congenital heart disease, with an estimated 2,400 babies affected in Australia each year. Most cases are diagnosed in infancy and early childhood, but diagnosis can also happen later in life. Ventricular septal defect is the most commonly reported congenital heart disease, followed by atrial septal defect and patent ductus arteriosus.

There were almost 5,000 congenital heart disease hospitalisations in 2016–17, with 1 in 3 among infants aged under 1 year, and 1 in 5 among children aged 1–14. Unlike other cardiovascular conditions, the number and rate of hospitalisation for congenital heart disease declines with age.

About half of all babies born with congenital heart disease will require surgical or catheter-based interventions at some time, with one-third needing these interventions in the first year of life. In 2016–17, there were over 2,700 surgical procedures conducted in Australian hospitals for closure of an atrial septal defect, ventricular septal defect or patent ductus arteriosus.

Congenital heart disease was the underlying cause of 152 deaths (0.1% of all deaths) in Australia in 2017. There were 70 deaths in infants aged under 1 year, which is equivalent to 6.9% of all infant deaths. Improvements in treatment and care in recent decades have led to more adults living with congenital heart disease. The decline in congenital heart disease mortality rates is reflected in an increase in the mean age of death of people with this disease.

There are several data gaps that prevent a comprehensive picture of the impact of congenital heart disease in Australia. Differences in collection and statistical methods across jurisdictions present difficulties in reporting national incidence and prevalence statistics. The *National Strategic Action Plan for Childhood Heart Disease 2019* recommends a framework for action and a range of activities regarding research and surveillance to set priorities and guide health policy and strategies, leading to improved health for people living with congenital heart disease.

1 Congenital heart disease in Australia

Childhood heart disease places a substantial burden on the health and wellbeing of children and their families, and on the Australian health-care system.

A number of different heart conditions can occur in childhood, with the distinction made between those that are present at birth (congenital heart disease) and those that develop later in childhood (acquired heart disease). This report looks at congenital heart disease.

Congenital heart disease is a common birth anomaly, estimated as affecting over 2,400 Australian babies each year. Around half require surgery or other interventions to manage their condition (Blue et al. 2012; Celemajer et al. 2016). Congenital heart disease is one of the leading causes of hospitalisation and death in infants.

A large number of children, young people and adults live with congenital heart disease. Many require complex, specialised care. However, improvements in treatment and care have extended survival and enable people with congenital heart disease to lead more active lives.

This report presents current national data on the number of people with congenital heart disease, on hospital treatment and care, and mortality.

What is congenital heart disease?

Congenital heart disease is a general term for any defect of the heart, heart valves or central blood vessels that is present at birth.

It can take many forms, including anomalies such as holes between the pumping chambers of the heart, valves that do not open or close properly and narrowing of major blood vessels such as the aorta and pulmonary artery (Box 1). Congenital heart disease can range from simple to complex, and more than 1 anomaly can occur in the same heart.

For moderate to complex congenital heart disease, diagnosis usually occurs within the first month of life. Common symptoms include bluish lips, fingers and toes, breathlessness or trouble breathing, low birth weight, difficulty feeding and gaining weight, and chest pain.

Although most cases are diagnosed in infancy and early childhood, diagnosis can happen later in life. Symptoms common in a later diagnosis may include abnormal heart rhythms, swelling, fainting, fatigue, abnormal breathing and dizziness.

Box 1: Selected congenital heart diseases

Ventricular septal defect—A hole in the muscle wall between the right and left ventricles.

Atrial septal defect—A hole in the muscle wall between the right and left atria.

Patent ductus arteriosus—Where the ductus arteriosus, the connection between the aorta and pulmonary artery, fails to close after birth.

Tetralogy of Fallot—A condition that consists of 4 heart anomalies: ventricular septal defect, a narrowing of the outflow tract into the pulmonary artery, an enlarged aorta and thickening of the muscle wall of the right ventricle.

Transposition of great vessels—A condition that is usually characterised by the aorta arising from the right ventricle and the pulmonary artery from the left ventricle.

(continued)

Box 1 (continued): Selected congenital heart diseases

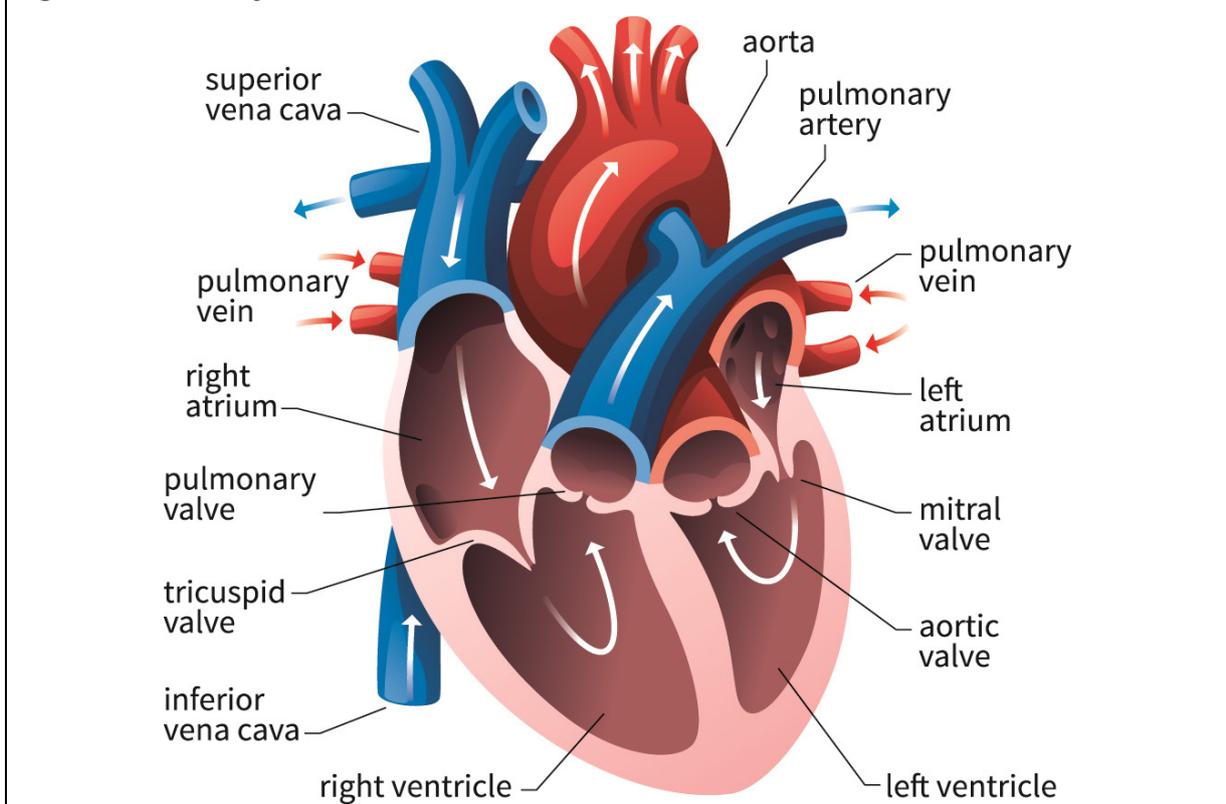
Coarctation of the aorta—Narrowing of the aorta.

Aortic stenosis—Obstruction of the aorta. This can be due to a narrowing of the aorta or a problem with the aortic valve.

Hypoplastic left heart syndrome—Where the left ventricle is small and functionally inadequate.

Pulmonary atresia—A condition in which there is no pulmonary valve and no blood flow to the pulmonary artery.

Figure 1: Anatomy of the heart



Causes of congenital heart disease

Most congenital heart disease is multifactorial and arises through combinations of genetic and environmental factors.

About one-third of cases can be attributed to chromosomal anomalies and gene disorders (Alankarage et al. 2018). Down syndrome is one of the most commonly seen syndromes in patients with congenital heart disease.

Some of the known risk factors include a family history of congenital heart disease, maternal illnesses such as rubella (German measles), misuse of alcohol, illicit drugs and over-the-counter or prescription medications, and maternal health factors such as early onset preeclampsia and poorly controlled diabetes (Blue et al. 2012; Jenkins et al. 2007).

2 How many people have congenital heart disease?

Globally, an estimated 9 in every 1,000 live births were affected by congenital heart disease during the period 2010–2017 (Liu et al. 2019).

National rates of congenital heart disease are not routinely reported. There are 5 states that currently publish the incidence of congenital heart disease. Data for New South Wales, Victoria, Western Australia and South Australia are summarised in Table 1.

Queensland congenital heart disease data are available from the Congenital Anomaly Linked File (CALF), which can be found on the Queensland Department of Health website (Queensland Department of Health 2018). These data are not comparable with data published by other jurisdictions due to the linkage methodology used to generate these statistics.

Table 1 reflects the number of congenital heart disease cases, and more than 1 case of congenital heart disease can be present in the same heart. The table should be interpreted with caution, because there are variations in data collection methods between jurisdictions. Of particular note are differences in reporting in the scope and severity of congenital anomalies, in inclusion of terminations of pregnancy, stillbirth and in the upper age limit of diagnosis, which ranges from 1 year in New South Wales to 6 years in Victoria and Western Australia.

Table 1: Reported number and rate of selected congenital heart disease cases, by state^(a)

	NSW	Vic	WA	SA
Congenital heart disease	2015	2015–2016	2014	2013
Number				
Transposition of great vessels	54	53	11	12
Tetralogy of Fallot	45	51	17	5
Ventricular septal defect	206	275	174	129
Atrial septal defect	208	44	50	34
Hypoplastic left heart syndrome	71		10	7
Patent ductus arteriosus	86	45	29	43
Coarctation of aorta	48	42	14	5
All cardiovascular defects^(b)	1,070	1,351	314	241
Number per 1,000 births				
Transposition of great vessels	0.6	0.3	0.3	0.6
Tetralogy of Fallot	0.5	0.3	0.5	0.2
Ventricular septal defect	2.1	1.7	5.3	6.4
Atrial septal defect	2.2	0.3	1.5	1.7
Hypoplastic left heart syndrome	0.7		0.3	0.3
Patent ductus arteriosus	0.9	0.3	0.9	2.1
Coarctation of aorta	0.5	0.3	0.4	0.2
All cardiovascular defects	11.1	8.5^(c)	9.5	11.9

(a) No recent data are available for Tasmania, ACT and NT. Queensland data are not comparable with data published by other jurisdictions.

(b) NSW, WA and SA: British Paediatric Association Classification of Diseases 74500–74799. Victoria: ICD-10 Q20–Q28.

(c) For Victoria, rates are number per 1,000 pregnancies.

Notes

1. The number of cardiovascular defects are reported—an individual can have more than 1 cardiovascular defect.
2. Data should be interpreted with caution, because there are variations in data collection methods between jurisdictions.

Sources: Gibson et al. 2017; NSW Ministry of Health 2019; Victorian Congenital Anomalies Register 2018; Western Australia Department of Health 2015.

Ventricular septal defect was the most commonly reported congenital heart disease, based on most recent reporting (784 cases in 4 states, noting 2 combined years of reporting for Victoria) followed by atrial septal defect (336) and patent ductus arteriosus (203).

Because the age for notification can range up to 6 years, there may be additional cases of congenital heart disease in some states that have not yet been reported.

Adults with congenital heart disease

Advances in paediatric cardiac care mean that people with congenital heart disease are now living longer, and the burden of disease is shifting from early childhood into the adult population (Celemajer et al. 2016).

Patients with complex and severe congenital heart disease will continue to require specialist treatment throughout their life. Often, they also require management of other health and welfare issues, including for family planning and pregnancy, lifestyle choices, dietary strategies, work choices and physical limitations.

In 2011, it was estimated that there were between 26,000 and 32,000 adults with congenital heart disease in Australia, with an annual increase of around 5% (Leggatt 2011).

Burden of disease and expenditure

Burden of disease analysis is useful to measure the impact of different diseases, conditions, injuries and risk factors on a population. It uses information from various sources to quantify the fatal and non-fatal effects of diseases.

Congenital heart disease is a leading contributor to the burden of disease among infants. The Australian Burden of Disease Study estimated that cardiovascular defects contributed 6.4% to the total disease burden of infants aged under 1 year in 2015 (AIHW 2019a). This burden fell by 29% between 2003 and 2015, from 8,120 to 5,730 disability adjusted life years (DALYs).

Cardiovascular defects also contribute to the burden of disease among older age groups, ranging from 1.0% of the burden for ages 1–4 to less than 0.1% for ages 65 and over.

An estimated \$199.5 million was spent on the diagnosis and treatment of cardiovascular defects in 2015–16, which is equivalent to 11% of recurrent expenditure on infant and congenital conditions and 0.2% of recurrent expenditure on all health conditions (AIHW 2019b). Just over half of this amount (\$105.7 million) was spent on infants aged under 1 year.

Most expenditure (82%) occurred among public hospital admitted patients, with another 10% for private hospital services.

3 Hospital treatment and care

Treatment of congenital heart disease has developed over recent decades, with improvements in diagnostic testing, surgical techniques and disease management.

Scanning for critical congenital heart disease is standard in Australia during second trimester ultrasound assessments. Early detection provides more time for planning and preparing for treatment after birth. Pulse oximetry and observation of vitals (such as Apgar) after birth while still in hospital are essential in both detecting and monitoring symptoms of congenital heart disease.

Treatments of congenital heart diseases are varied. There are many medications as well as surgical and catheter-based treatments that manage oxygen levels and blood pressure and repair physical anomalies. Treatment usually begins in infancy or early childhood.

A summary of common diagnostic and treatment options for congenital heart disease is shown in Box 2.

Box 2: Diagnosis and treatment of congenital heart disease

Echocardiography—An ultrasound of the heart. This test is non-invasive and can be conducted before birth.

Pulse oximetry—A non-invasive test that measures the oxygen levels in the blood to see how efficiently the heart is pumping oxygen to the rest of the body.

Apgar—An observational test done at 1 minute and then 5 minutes after birth that checks skin colour, heart rate, responsiveness, muscle tone and breathing rate.

Medications—Often used for mild congenital heart defects, especially those found later in childhood or in adulthood. Medications can help the heart work more efficiently by lowering blood pressure, regulating the heartbeat or lowering the amount of fluid in the chest.

Cardiac catheterisation—A thin flexible tube is inserted into an artery in the leg and moved towards the heart to measure blood pressure and flow. Sometimes used in conjunction with imaging procedures including contrast studies and X-ray. Also a form of treatment to stretch narrowed vessels and valves, implant stents or close a hole.

Corrective surgery—Usually reserved for more complex congenital heart conditions. There are many different procedures. Surgery is often undertaken in the first year of life.

Heart transplant—Total replacement of the heart muscle.

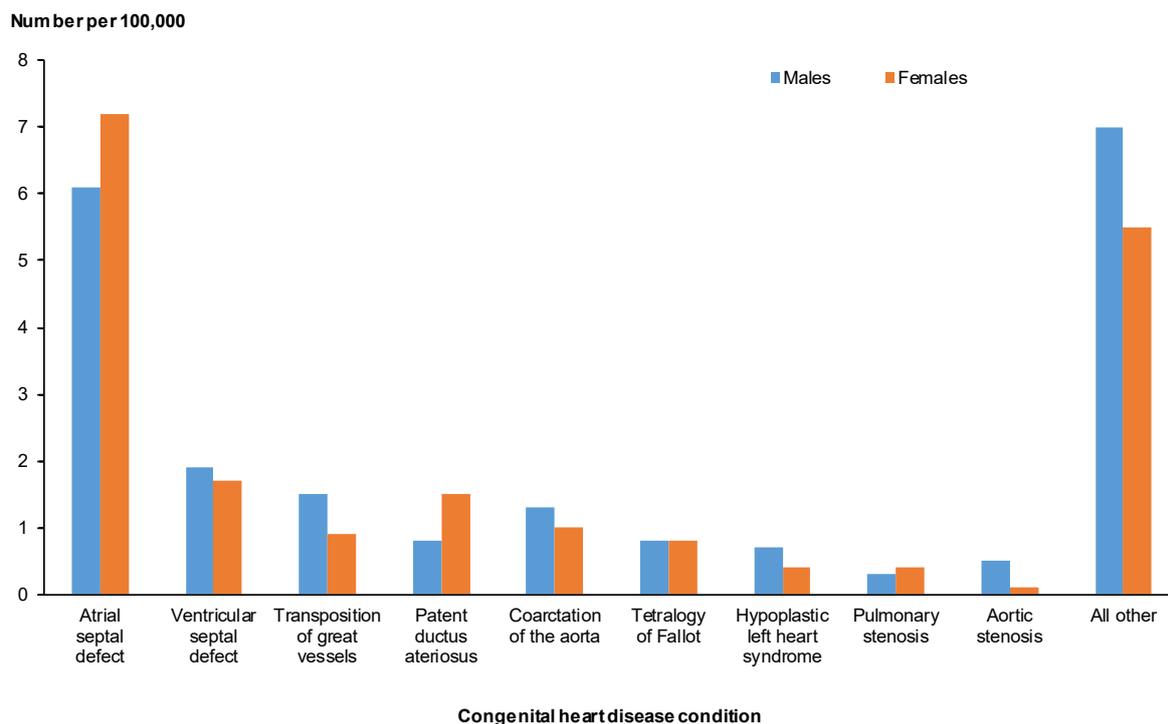
Compassionate care—An alternative to surgery, often using palliation and other forms of end-of-life care.

In 2016–17, there were around 4,900 hospitalisations in Australia where congenital heart disease was the principal diagnosis—a rate of 20 hospitalisations per 100,000 population.

The highest rate of hospitalisation for a specific form of congenital heart disease was for atrial septal defect (6.6 hospitalisations per 100,000 population), followed by ventricular septal defect (1.8), transposition of the great vessels (1.2), patent ductus arteriosus and coarctation of the aorta (both 1.1) (Figure 2).

Note that these data are based on ‘episodes of care’ rather than the number of people hospitalised with a condition. A person might be hospitalised more than once in any year.

Figure 2: Congenital heart disease hospitalisations, principal diagnosis, by condition and sex, 2016–17



Note: Age-standardised to the 2001 Australian population.

Source: AIHW National Hospital Morbidity Database. See Table S1.

Having congenital heart disease can also impact on the treatment and care received by persons who are hospitalised for other reasons.

In 2016–17, there were more than 10,000 hospitalisations across all age groups where congenital heart disease was an additional diagnosis. For these cases, the principal diagnoses—the diagnosis chiefly responsible for the hospitalisation—included:

- diseases of the circulatory system, such as non-rheumatic aortic valve disorders and cerebral infarction; 2,900 (29%)
- certain conditions originating in the perinatal period, such as disorders related to short gestation and low birth weight, not elsewhere classified; 2,700 (27%)
- congenital malformations, deformations and chromosomal abnormalities; 2,100 (21%)
- symptoms, signs and abnormal clinical and laboratory findings, not elsewhere classified, such as pain in throat and chest and abnormalities breathing; 522 (5.2%)
- diseases of the respiratory system; 505 (5.0%).

Sex and age

In 2016–17, the hospitalisation rate for congenital heart disease was similar for males and females (21 and 20 per 100,000, respectively).

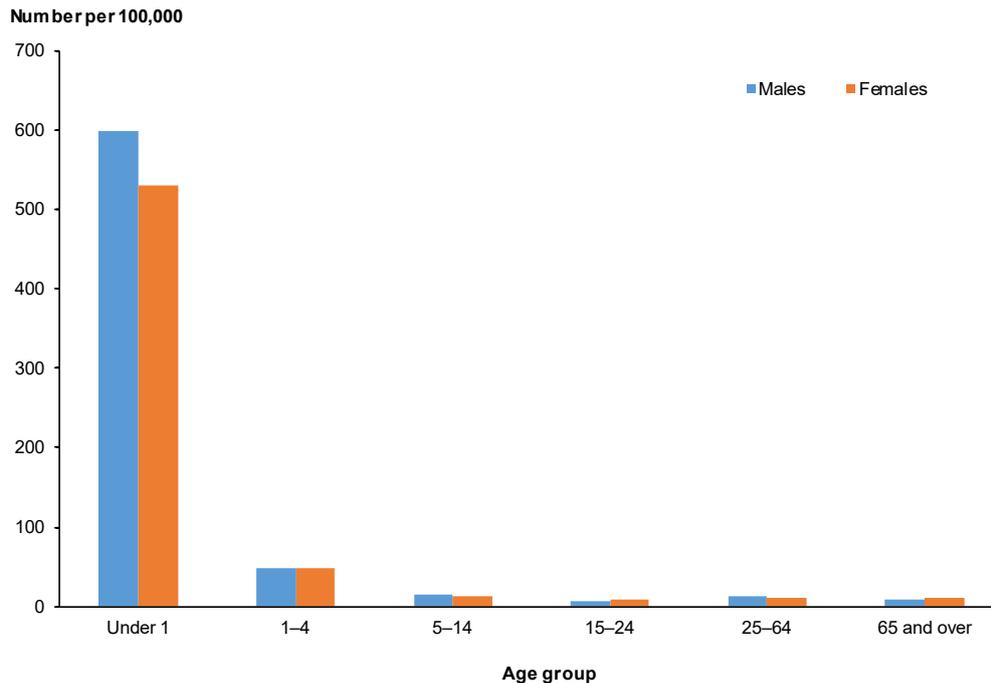
Unlike other cardiovascular conditions, the number and rate of hospitalisation for congenital heart disease declines with age.

One in 3 hospitalisations for congenital heart disease (1,700 or 36%) were for infants aged under 1 year. The rate for infant boys (599 per 100,000) was higher than for infant girls (530 per 100,000) (Figure 3).

One in 5 hospitalisations (1,050 or 22%) were among children aged 1–14.

Hospitalisation for congenital heart disease, however, can occur at any age. Almost half of all congenital heart disease hospitalisations in 2016–17 were persons aged 15 and over, with 4.9% (240) aged 15–24, one-third (1,500 or 31%) aged 25–64 and 7.1% (340) aged 65 and over.

Figure 3: Congenital heart disease hospitalisations, by age and sex, 2016–17



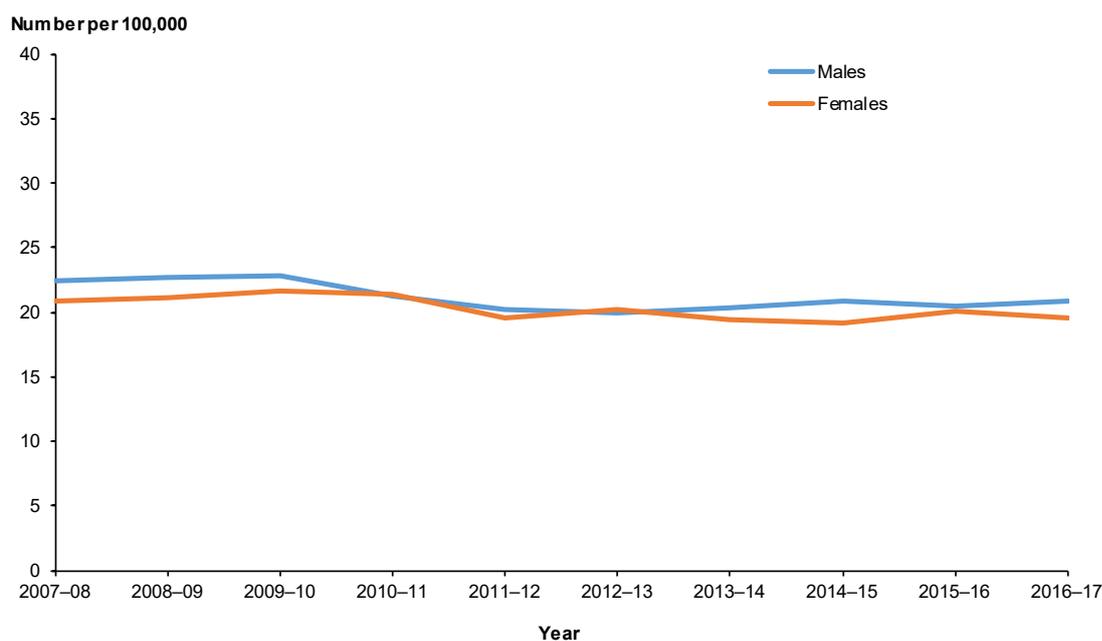
Source: AIHW National Hospital Morbidity Database. See Table S3.

Trends

Between 2007–08 and 2016–17, the hospitalisation rate for congenital heart disease decreased by 6.5% from 22 to 20 per 100,000 population. Trends in rate were similar for male and females (Figure 4).

Trends in rates of hospitalisation for earlier years had shown increases: between 1993–94 and 2007–08, the age-standardised hospitalisation rate for all cardiovascular defects increased by 13% from 24 to 28 per 100,000 population. This increase may reflect changes in diagnostic technology over this period, including for fetal cardiac diagnosis (AIHW 2011; Leggatt 2011).

Figure 4: Congenital heart disease hospitalisations, by sex, 2007–08 to 2016–17



Note: Age-standardised to the 2001 Australian population.

Source: AIHW National Hospital Morbidity Database. See Table S4.

Population groups

Aboriginal and Torres Strait Islander people

In 2016–17, there were 249 hospitalisations of Aboriginal and Torres Strait Islander people with congenital heart disease. There was no statistically significant difference between Indigenous and non-Indigenous hospitalisation rates for congenital heart disease (22 and 20 per 100,000 population, respectively).

Remoteness

In 2016–17, there was no statistically significant difference in congenital heart disease hospitalisation rates between *Major cities*, *Regional* and *Remote* areas (20, 21 and 23 per 100,000 population, respectively).

Socioeconomic position

In 2016–17, the congenital heart disease hospitalisation rate in the lowest socioeconomic group (21 per 100,000 population) was higher than in the highest socioeconomic group (17 per 100,000 population).

Length of stay

Congenital heart disease has a long average length of hospital stay.

Among those with congenital heart disease hospitalised for more than 1 night in 2016–17, the average length of stay was 9.0 days (9.5 days for males and 8.5 days for females).

The average length of stay has varied slightly over the last decade, from a low of 8.0 days in 2008–09 to a high of 9.2 days in 2015–16.

Other cardiovascular diseases with long average lengths of stay include cerebrovascular diseases (stroke) (11.5 days in 2016–17) and chronic rheumatic heart disease, an acquired childhood heart disease (11.4 days).

Procedures

About half of all babies born with congenital heart disease require surgical or catheter-based interventions to correct any defect, with one-third needing these interventions in the first year of life (Blue et al. 2012; Leggatt 2011).

Where congenital heart disease was the principal or additional diagnosis, there were over 1,600 surgical procedures conducted in Australian hospitals for closure of an atrial septal defect in 2016–17, around 440 for closure of ventricular septal defect and 650 for closure of patent ductus arteriosus.

Procedure rates for infants aged under 1 year were 105 per 100,000 for atrial septal defect, 99 per 100,000 for ventricular septal defect and 124 per 100,000 for patent ductus arteriosus in 2016–17.

Where procedures were performed for patent ductus arteriosus and ventricular septal defect, most were among children aged under 5 (74% and 83%, respectively).

Procedures for atrial septal defect were spread more evenly over all ages, with only 27% of procedures in children aged under 5, and 46% in adults aged 45 and over.

Trends in procedure rates have changed little over the last decade—for atrial septal defect (6.2 and 6.6 per 100,000 in 2007–08 and 2016–17, respectively), ventricular septal defect (2.1 and 1.8 per 100,000 in 2007–08 and 2016–17, respectively) and patent ductus arteriosus (2.8 and 2.7 per 100,000 in 2007–08 and 2016–17, respectively).

Deaths in hospital

Death from congenital heart disease while in hospital is a comparatively rare event.

Of hospitalisations in 2016–17 with a principal diagnosis of congenital heart disease, 0.8% had a separation mode of 'died', which was down from 1.3% in 2007–08.

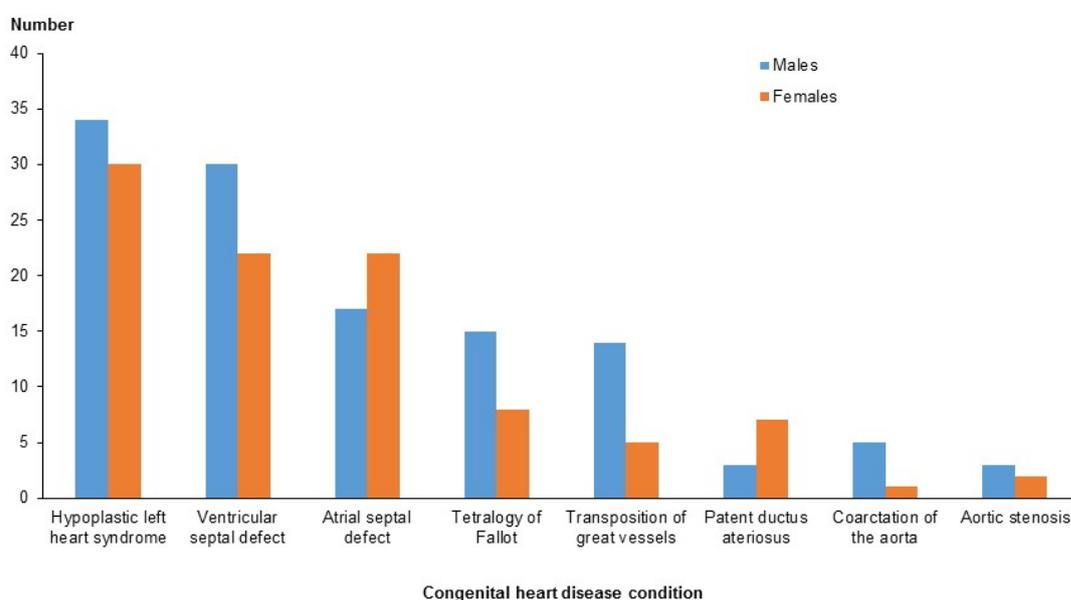
4 Deaths

In 2017, congenital heart disease was the underlying cause of 152 deaths (0.1% of all deaths) in Australia.

Congenital heart disease is a leading cause of death among infants. In 2017, congenital heart disease caused 70 deaths in infants aged under 1 year, equivalent to 6.9% of all infant deaths.

Among 469 congenital heart disease deaths that occurred among all ages during the 3 years 2015–2017, hypoplastic left heart syndrome resulted in the highest number of deaths (64), followed by ventricular septal defect (52), atrial septal defect (39), Tetralogy of Fallot (23) and transposition of the great vessels (19) (Figure 5).

Figure 5: Congenital heart disease deaths, underlying cause, by condition and sex, 2015–2017



Source: AIHW National Mortality Database. See Table S14.

Congenital heart disease deaths also comprise a substantial proportion of perinatal deaths. Perinatal deaths refer to stillbirths (fetal deaths) and deaths of infants within the first 28 days of life (neonatal deaths).

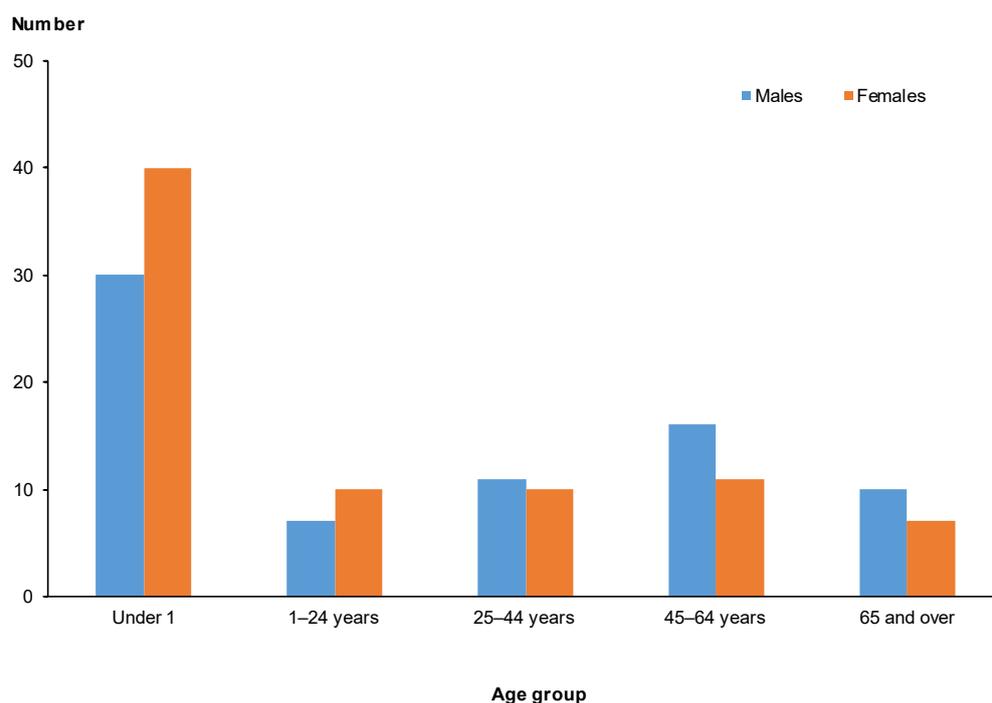
In 2017, there were 119 perinatal deaths with congenital heart disease as the main condition, of which 70 were fetal deaths and 49 were neonatal deaths. Congenital heart disease was responsible for 4.8% of all perinatal deaths in 2017 (ABS 2018).

Sex and age

In 2017, 74 males and 78 females died as a result of congenital heart disease.

Forty-six per cent were aged 1 year or under (30 boys and 40 girls), 11% (17) were aged 1–24, 14% (21) were aged 25–44, 18% (27) were aged 45–64 and 11% (17) were aged 65 or over (Figure 6).

Figure 6: Congenital heart disease deaths, underlying cause, by age and sex, 2017



Source: AIHW National Mortality Database. See Table S15.

Trends

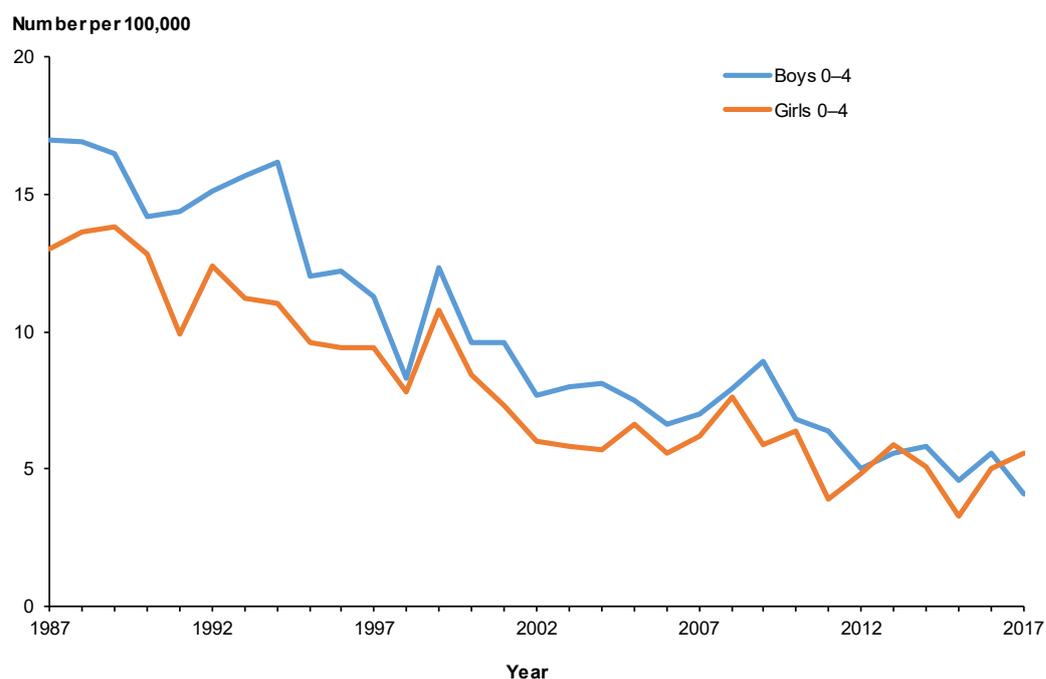
The age-standardised death rate for males fell from 1.7 deaths per 100,000 in 1987 to 0.6 in 2017, and for females from 1.2 in 1987 to 0.6 per 100,000 in 2017.

Fewer deaths as a result of treatment improvements and an increase in the number of terminations of pregnancies after antenatal diagnosis have been suggested as factors that have contributed to the decrease in mortality rates (Leggatt 2011).

The age-specific congenital heart disease death rate for children under 5 years also fell—from 15.1 deaths per 100,000 population in 1987, to 4.8 in 2017 (Figure 7).

In 1987, 71% of congenital heart disease deaths occurred in children aged under 5. By 1997, this figure had fallen to 55%, falling further to 50% in 2007 and remained at 50% in 2017.

Figure 7: Congenital heart disease death rates for children aged under 5, by sex, 1987 to 2017

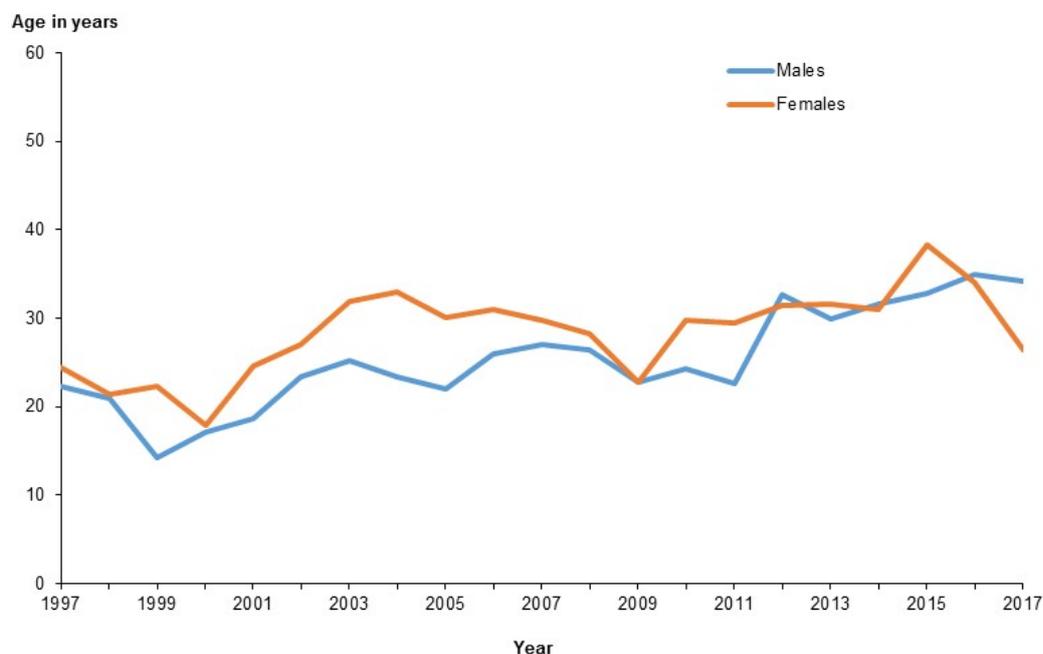


Source: AIHW National Mortality Database. See Table S17.

People with congenital heart disease are, on average, living longer. The decline in congenital heart disease mortality rates is reflected in an increase in the mean age of death of persons, where congenital heart disease was either an underlying or associated cause of death (Figure 8).

The mean age of death for these persons increased from 23.3 years in 1997, to 28.2 years in 2007 and 30.4 years in 2017.

Figure 8: Mean age at death where congenital heart disease is either an underlying or associated cause of death, 1997 to 2017



Source: AIHW National Mortality Database. See Table S18.

Population groups

Aboriginal and Torres Strait Islander people

In 2015–2017, congenital heart disease was the cause of death of 29 Aboriginal and Torres Strait Islander people in the 5 jurisdictions with adequate Indigenous identification (see Appendix A).

There was no statistically significant difference in congenital heart disease death rates between Indigenous and non-Indigenous people in these jurisdictions (1.1 and 0.6 per 100,000 population, respectively).

Remoteness

In 2015–2017, there was no statistically significant difference in congenital heart disease death rates between *Major cities*, *Regional* and *Remote* areas (0.6, 0.8 and 0.7 per 100,000 population, respectively).

Socioeconomic position

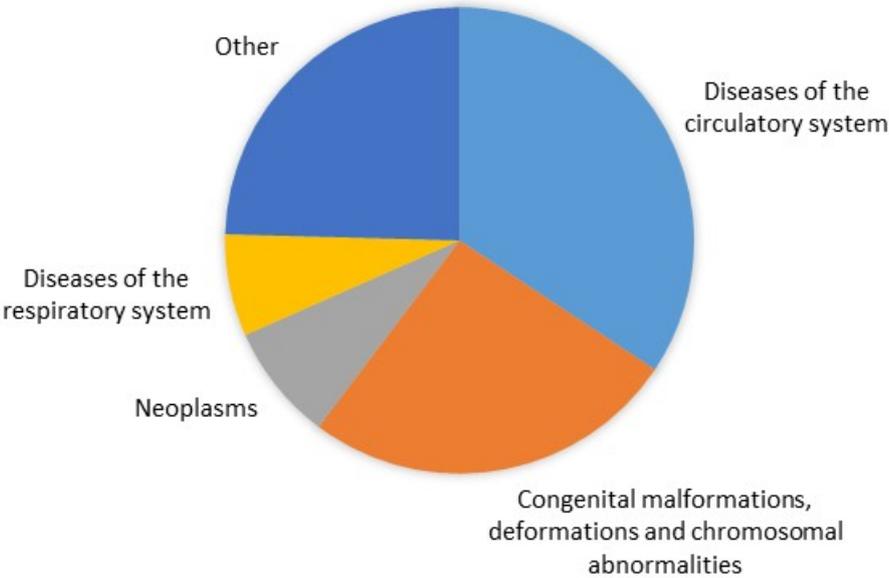
In 2015–2017, the congenital heart disease death rate in the lowest socioeconomic group (0.9 per 100,000 population) was higher than in the highest socioeconomic group (0.4).

Associated cause of death

A more complete picture of the mortality burden of congenital heart disease can be obtained by examining the underlying cause of death when congenital heart disease is listed on a death certificate as an associated cause of death.

In 2015–2017, congenital heart disease was listed as an associated cause of death on 298 death certificates. The most common underlying causes in these deaths were diseases of the circulatory system (34%), congenital malformations, deformations and chromosomal abnormalities (26%), neoplasms (8.1%) and diseases of the respiratory system (7.0%) (Figure 9).

Figure 9: Most common underlying causes of death where congenital heart disease is an associated cause, 2015–2017



Source: AIHW National Mortality Database. See Table S20.

5 Data gaps

There are a number of data issues that hinder the formation of a comprehensive picture of the impact of congenital heart disease in Australia. Differences in definitions and in data collection methods between jurisdictions, for example, leads to difficulties in reporting the national incidence and prevalence of congenital heart disease.

The Congenital Heart Alliance of Australia and New Zealand (CHAANZ), established in 2014, is seeking to develop an Australian and New Zealand congenital heart disease registry to inform the future needs of patients and healthcare services. CHAANZ conducts the National Congenital Heart Disease Survey, a voluntary self-reported questionnaire on patient experience and needs of those with congenital heart disease (CHAANZ 2019).

The National Strategic Action Plan on Childhood Heart Disease outlines areas relating to surveillance, and the collection and analysis of data, where improvements could lead to better care for people living with congenital heart disease (Department of Health 2019).

Surveillance activities identified in the Action Plan include:

- using the CHAANZ registry and national survey to track the impact of Action Plan recommendations over time
- conducting focus group interviews of key stakeholders and implementation partners
- undertaking economic impact reports to measure any improvements to the economic and productivity burden of the disease on the Australian community
- publishing an Action Plan annual report card.

Data linkage and improved integration of health data sets can provide a more robust picture of the use of health services among people with congenital heart disease, and greater evidence for causal pathways to good health.

Appendix: Data sources and classifications

AIHW National Hospital Morbidity Database

The AIHW National Hospital Morbidity Database (NHMD) is a compilation of episode-level records from admitted patient morbidity data collection systems in Australian hospitals. The data supplied are based on the National minimum data set (NMDS) for admitted patient care and include demographic, administrative and length of stay data, as well as data on the diagnoses of the patients, the procedures they underwent in hospital and external causes of injury and poisoning.

The purpose of the NMDS for Admitted patient care is to collect information about care provided to admitted patients in Australian hospitals. The scope of the NMDS is episodes of care for admitted patients in all public and private acute and psychiatric hospitals, free-standing day hospital facilities, and alcohol and drug treatment centres in Australia. Hospitals operated by the Australian Defence Force, corrections authorities and in Australia's off-shore territories are not in scope but some are included.

The counting unit in the NHMD is the 'separation', described as 'hospitalisations' in this report. 'Separation' is the term used to refer to the episode of admitted patient care, which can be a total hospital stay (from admission to discharge, transfer or death) or a portion of a hospital stay beginning or ending in a change of type of care (for example, from acute care to rehabilitation).

The hospital separations data do not include episodes of non-admitted patient care provided in outpatient clinics or emergency departments. Patients in these settings may be admitted subsequently, with the care provided to them as admitted patients being included in the NHMD. The following care types were excluded from analysis: 7.3 (newborn—unqualified days only), 9 (organ procurement—posthumous) and 10 (hospital border).

Data on diagnoses are recorded uniformly using the 9th edition of the *International Statistical Classification of Diseases and Related Health Problems, 10th Revision, Australian Modification* (ICD-10-AM) (ACCD 2014).

Further information about the NHMD can be found in the Data quality statement: National Hospital Morbidity Database 2014–15 at <http://meteor.aihw.gov.au/content/index.phtml/itemId/638202>.

AIHW National Mortality Database

The AIHW National Mortality Database (NMD) contains information provided by the registries of births, deaths and marriages and the National Coronial Information System (coded by the ABS), for deaths from 1964 to 2017. Registration of deaths is the responsibility of each state and territory's registry of births, deaths and marriages. These data are then collated and coded by the ABS and maintained at the AIHW in the NMD.

In this report, deaths registered in 2014 and earlier are based on the final version of cause of death data; deaths registered in 2015 are based on revised version; and deaths registered in 2016 and 2017 are based on preliminary version. Revised and preliminary versions are subject to further revision by the ABS.

Mortality data by Indigenous status are reported for 5 jurisdictions combined: New South Wales, Queensland, Western Australia, South Australia and the Northern Territory. The

AIHW considers the quality of Indigenous identification in mortality data for these 5 jurisdictions to be adequate from 1998.

The data quality statements underpinning the AIHW NMD can be found on the following ABS internet pages:

- ABS quality declaration summary for *Deaths, Australia* (ABS cat. no. 3302.0)
<http://www.abs.gov.au/ausstats/abs%40.nsf/mf/3302.0/>
- ABS quality declaration summary for *Causes of death, Australia* (ABS cat. no. 3303.0)
<http://www.abs.gov.au/ausstats/abs%40.nsf/mf/3303.0/>.

For more information on the AIHW NMD and deaths data, see

<https://www.aihw.gov.au/about-our-data/our-data-collections/national-mortality-database/deaths-data>.

Australian Burden of Disease Study 2015

Data to develop the burden of disease estimates In the Australian Burden of Disease Study 2015 were obtained from numerous sources. Deaths data used to estimate fatal burden were sourced from the NMD. Morbidity data used to estimate non-fatal burden were drawn from disease registers, administrative data, surveys and epidemiological studies.

Risk factor exposure data were sourced from national surveys, registry data and monitoring programs. Relative risks were obtained from the Global Burden of Disease Study 2016 and from an AIHW review of literature.

Other inputs, such as the standard life table for fatal burden, and the health states and disability weights for non-fatal burden, were obtained from the Global Burden of Disease Study 2010, 2013 or 2015. Population estimates were sourced from the ABS.

For full details on methods, data sources and standard inputs see *Australian Burden of Disease Study: methods and supplementary material 2015* (AIHW 2019c), available on the AIHW website at <https://www.aihw.gov.au/reports-statistics/health-conditions-disability-deaths/burden-of-disease/reports>.

Disease classifications

Table A1: ICD-10-AM (9th edition) codes used to define conditions in hospitalisations data

Condition	ICD-10-AM codes
Congenital heart disease	Q20–Q26
Transposition of great vessels	Q20.1, Q20.3, Q20.5
Ventricular septal defect	Q21.0
Atrial septal defect	Q21.1
Tetralogy of Fallot	Q21.3
Congenital pulmonary valve stenosis	Q22.1
Congenital stenosis and atresia of aortic valve	Q23.0
Hypoplastic left heart syndrome	Q23.4
Patent ductus arteriosus	Q25.0
Coarctation of the aorta	Q25.1

Source: ACCD 2014.

Table A2: ICD-10 codes used to define deaths

Condition	ICD-10 codes
Congenital heart disease	Q20–Q26
Transposition of great vessels	Q20.1, Q20.3, Q20.5
Ventricular septal defect	Q21.0
Atrial septal defect	Q21.1
Tetralogy of Fallot	Q21.3
Congenital pulmonary valve stenosis	Q22.1
Congenital stenosis of aortic valve	Q23.0
Hypoplastic left heart syndrome	Q23.4
Patent ductus arteriosus	Q25.0
Coarctation of aorta	Q25.1

Note: Congenital heart disease deaths occurring before 1997 were classified to ICD-9 codes 745–746 and adjusted with a comparability factor of 1.12.

Source: WHO 2016.

Table A3: ACHI (9th edition) codes used to define procedures

Procedure codes	ACHI codes
Atrial septal defect	Block: 617
Ventricular septal defect	Block: 618
Patent ductus arteriosus	Block: 690

Source: ACCD 2015.

More information

HeartKids provides advocacy, information, research and support for children, young people and adults living with congenital heart disease: <https://www.heartkids.org.au/>.

The National Strategic Action Plan for Childhood Heart Disease 2019 presents an overview of Australian childhood heart disease and identifies issues and potential areas for future actions (Department of Health 2019).

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Abbreviations

ABS	Australian Bureau of Statistics
ACCD	Australian Consortium for Classification Development
ACHI	Australian Classification of Health Interventions
AIHW	Australian Institute of Health and Welfare
CHAANZ	Congenital Heart Alliance of Australia and New Zealand
DALYs	disability-adjusted life years
ICD-10	International Statistical Classification of Diseases and Related Health Problems, 10th revision
ICD-10-AM	International Statistical Classification of Diseases and Related Health Problems, 10th revision, Australian Modification
NHMD	National Hospital Morbidity Database
NMD	National Mortality Database
NMDS	National minimum data set
WHO	World Health Organization

Glossary

additional diagnosis: A condition or complaint either coexisting with the principal diagnosis or arising during the episode of admitted patient care, episode of residential care or attendance at a health-care establishment.

admitted patient: A patient who undergoes a hospital's admission process to receive treatment and/or care. This treatment and/or care is provided over a period of time and can occur in hospital and/or in the person's home.

age-standardisation: A set of techniques used to remove as far as possible the effects of differences in age when comparing 2 or more populations.

antenatal: The period covering conception up to the time of birth. Synonymous with prenatal.

Australian Classification of Health Interventions (ACHI): The procedure classification is divided into subchapters by anatomical site, which are further divided into more specific procedure blocks, beginning with the least invasive procedure through to the most invasive. The blocks, which are numbered sequentially, group specific procedure codes.

average length of stay: The average number of patient days for admitted patient episodes. Patients admitted and separated on the same day are allocated a length of stay of 1 day.

associated cause(s) of death: A cause(s) listed on the Medical Certificate of Cause of Death, other than the underlying cause of death. They include the immediate cause, any intervening causes, and conditions that contributed to the death but were not related to the disease or condition causing death.

care type: The care type defines the overall nature of a clinical service provided to an admitted patient during an episode of care (admitted care), or the type of service provided by the hospital for boarders or posthumous organ procurement (care other than admitted care).

cause(s) of death: All diseases, morbid conditions or injuries that either resulted in or contributed to death—and the circumstances of the accident or violence that produced any such injuries—that are entered on the Medical Certificate of Cause of Death. Causes of death are commonly reported by the underlying cause of death.

congenital: A condition that is recognised at birth, or is believed to have been present since birth, including conditions inherited or caused by environmental factors.

disability-adjusted life years: A measure of healthy life lost, either through premature death or living with disability due to illness or injury.

episode of care: The period of admitted patient care between a formal or statistical admission and a formal or statistical separation, characterised by only 1 care type.

fetal death (stillbirth): Death, before the complete expulsion or extraction from its mother, of a product of conception of 20 or more completed weeks of gestation, or of 400 g or more birthweight. Death is evidenced by the fact that, after such separation, the fetus does not breathe or show any other signs of life, such as beating of the heart, pulsation of the umbilical cord or definite movement of voluntary muscles.

hospitalisation: Synonymous with separation; that is, an episode of hospital care that starts with the formal admission process and ends with the formal separation process (see **separation**).

Indigenous: a person of Aboriginal and/or Torres Strait Islander descent who identifies as an Aboriginal and/or Torres Strait Islander.

International Classification of Diseases (ICD): The World Health Organization's internationally accepted classification of diseases and related health conditions.

multiple causes of death: All the causes listed on the Medical Certificate of Cause of Death. These include the underlying cause of death and all associated cause(s) of death.

neonatal death: The death of a live born baby within 28 days of birth.

paediatric: Relating to the branch of medicine dealing with children and their diseases.

perinatal: Describes something that pertains to, or that occurred in, the period shortly before or after birth (usually up to 28 days after).

prevalence: The number or proportion (of cases, instances, and so forth) in a population at a given time.

principal diagnosis: The diagnosis established after study to be chiefly responsible for occasioning an episode of admitted patient care, an episode of residential care or an attendance at the health-care establishment.

procedure: A clinical intervention that is surgical in nature, carries a procedural risk, carries an anaesthetic risk, requires specialised training and/or requires special facilities or equipment available only in the acute care setting.

remoteness area: A classification of the remoteness of a location using the Australian Statistical Geography Standard Remoteness Structure (2011), based on the Accessibility /Remoteness Index of Australia (ARIA), which measures the remoteness of a point based on the physical road distance to the nearest urban centre. The categories are: *Major cities, Inner regional, Outer regional, Remote, Very remote* and *Migratory*.

trimester: Division of human pregnancy, which is frequently divided into 3 terms of about 3 months each to refer to the fetus's development.

separation: An episode of care for an admitted patient, which can be a total hospital stay (from admission to discharge, transfer or death), or a portion of a hospital stay beginning or ending in a change of type of care (for example, from acute to rehabilitation). Separation also means the process by which an admitted patient completes an episode of care either by being discharged, dying, transferring to another hospital or changing type of care.

socioeconomic position: An indication of how 'well off' a person or group is. In this report, socioeconomic areas are reported using the Socio-Economic Indexes for Areas, for 5 groups (quintiles)—from the most disadvantaged (worst off or lowest socioeconomic area) to the least disadvantaged (best off or highest socioeconomic area).

underlying cause of death: The disease or injury that initiated the train of events leading directly to death, or the circumstances of the accident or violence that produced the fatal injury.

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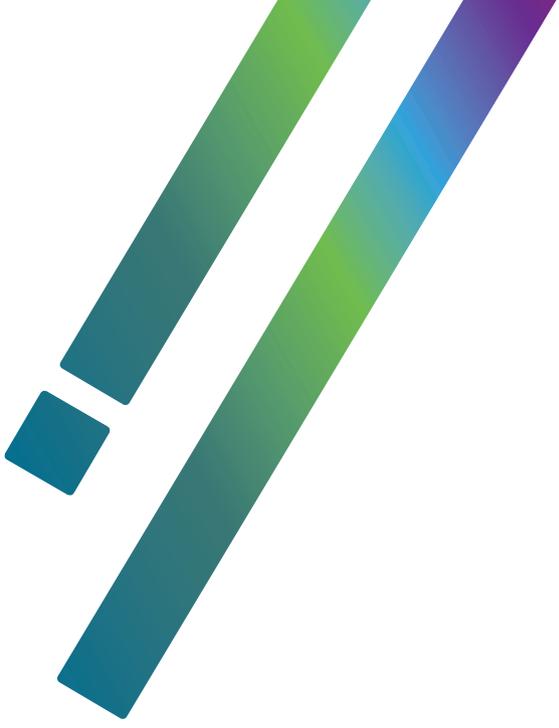
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Globally, congenital heart disease affects around 9 in every 1,000 babies born. In Australia, a large number of children, young people and adults live with congenital heart disease. This report presents latest statistics and trends on the incidence of congenital heart disease, and on hospitalisation and mortality.

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