



American  
Heart  
Association.

## 2022 Heart Disease & Stroke Statistical Update Fact Sheet Congenital Cardiovascular Defects

Congenital cardiovascular defects (CCD) arise from abnormal or incomplete formation of the heart and blood vessels. CCDs range in severity from minor abnormalities not requiring treatment to complex malformations, including absent, hypoplastic, or atretic portions of the heart, valves, or vessels that could require multiple surgeries and interventions, including cardiac transplantation. Thus, there is significant variability in their presentation and requirements for care that can have a significant impact on morbidity, mortality, and healthcare costs both in children and adults.

The National Birth Defects Prevention Network showed the average birth prevalence of 21 selected major birth defects for 13 states in the United States from 2004 to 2006. These data indicated that there are >6100 estimated annual cases of 5 CCDs: truncus arteriosus (0.07 per 1000 births), TGA (0.3 per 1000 births), tetralogy of fallot (TOF) (0.4 per 1000 births), atrioventricular septal defect (0.47 per 1000 births), and HLHS (0.23 per 1000 births).

### Incidence

- An estimated minimum of 40,000 infants are expected to be affected each year by congenital heart defects in the United States. Of these, about 25%, or 2.4 per 1,000 live births, require invasive treatment in the first year of life.
- In high-income North America, including the United States, the birth prevalence of CCDs is estimated to be 12.3 per 1000.

### Prevalence

- The 2017, the all-age prevalence of CCDs in the United States was estimated at 466,566 individuals, with 279,320 (60%) of these <20 years of age.

### Mortality

- Mortality related to congenital cardiovascular defects in 2019 was 2,890 for all ages.
- In 2019, the age-adjusted death rate attributable to CCDs was 0.9 death per 100,000 people, an 18.2% decrease from 2009.
- In 2019, CCDs were the most common causes of infant death resulting from birth defects; 21.6% of infants who died in 2019 of a birth defect had a heart defect.

### Risk Factors

- Numerous intrinsic and extrinsic nongenetic risk factors contribute to congenital heart defects.
- Twins are at higher risk for congenital heart defects.
- Known maternal risks include maternal smoking during the first trimester of pregnancy.
- Exposure to secondhand smoke has also been implicated as a risk factor.
- Exposure to air pollution increases risk.
- Maternal binge drinking is also associated with an increased risk of congenital cardiac defects, and the combination of binge drinking and smoking may be particularly dangerous.
- Maternal obesity is associated with congenital heart defects.

## Risk Factors (continued)

- Maternal diabetes, including gestational diabetes, has also been associated with cardiac defects, both isolated and multiple. Pre-gestational diabetes is also associated with congenital heart defects, specifically TOF.
- Folate deficiency is a well-documented risk for congenital malformations, including congenital heart defects, and folic acid supplementation is recommended during pregnancy.
- Maternal infections, including rubella and chlamydia, have been associated with congenital heart defects.
- Paternal exposures that increase risk for congenital heart defects include paternal anesthesia, sympathomimetic medication, pesticides, solvents and phthalates.

## Hospitalizations & Costs

- Among pediatric hospitalizations (age 0–20 years) in 2012:
  - Pediatric hospitalizations with congenital heart defects (4.4% of total pediatric hospitalizations) accounted for \$6.6 billion in hospitalization spending (23% of total pediatric hospitalization costs).
  - 26.7% of all congenital heart defect costs were attributed to critical congenital heart defects, with the highest costs attributable to hypoplastic left heart syndrome (HLHS), coarctation of the aorta, and TOF.
  - Mean cost of congenital heart defects was higher in infancy (\$36,601) than in older ages and in those with critical congenital heart defects (\$52,899).

**For additional information, charts and tables, see**

***Heart Disease & Stroke Statistics – 2022 Update***

Additional charts may be downloaded directly from the online publication or [www.heart.org/statistics](http://www.heart.org/statistics).

Many statistics in this At-a-Glance document come from unpublished tabulations compiled for this document and can be cited using the document citation listed below. The data sources used for the tabulations are listed in the full document. Additionally, some statistics come from published studies. If you are citing any of the statistics in this At-a-Glance document, please review the full Heart Disease and Stroke Statistics document to determine data sources and original citations.

The American Heart Association requests that this document be cited as follows:

Tsao CW, Aday AW, Almarzoq ZI, Alonso A, Beaton AZ, Bittencourt MS, Boehme AK, Buxton AE, Carson AP, Commodore-Mensah Y, Elkind MSV, Evenson KR, Eze-Nliam C, Ferguson JF, Generoso G, Ho JE, Kalani R, Khan SS, Kissela BM, Knutson KL, Levine DA, Lewis TT, Liu J, Loop MS, Ma J, Mussolino ME, Navaneethan SD, Perak AM, Poudel R, Rezk-Hanna M, Roth GA, Schroeder EB, Shah SH, Thacker EL, VanWagner LB, Virani SS, Voecks JH, Wang N-Y, Yaffe K, Martin SS; on behalf of the American Heart Association Council on Epidemiology and Prevention Statistics Committee and Stroke Statistics Subcommittee. Heart disease and stroke statistics—2022 update: a report from the American Heart Association [published online ahead of print Wednesday, January 26, 2022]. *Circulation*. doi: 10.1161/CIR.0000000000001052

Link to document: <https://www.ahajournals.org/doi/10.1161/CIR.0000000000001052>

If you have questions about statistics or any points made in the Statistical Update, please contact the American Heart Association National Center, Office of Science & Medicine at [statistics@heart.org](mailto:statistics@heart.org). Please direct all media inquiries to News Media Relations at <http://newsroom.heart.org/newsmedia/contacts>.