



American
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2021 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 most common complex congenital cardiovascular defects* and percent distribution, among adults and children, include the following:

- Ventricular septal defect (VSD) – 20.1%
- Atrial septal defect (ASD) – 18.8%
- Patent ductus arteriosus – 14.2%
- Valvular pulmonic stenosis – 13.5%
- Coarctation of the aorta – 7.6%
- Tetralogy of Fallot (TOF) – 6.1%
- Valvular aortic stenosis – 5.4%
- Atrioventricular septal defect – 3.1%
- Transposition of the great arteries (TGA) – 2.6%
- Hypoplastic right heart syndrome – 2.2%

*2002 U.S. prevalence data; excludes an estimated 3 million bicuspid aortic valve prevalence (2 million in adults and 1 million in children).

Incidence

- Variations in birth prevalence of congenital heart defects have been reported from 6.9 per 1000 live births in North America, 8.2 per 1000 in Europe, and 9.3 per 1000 in Asia.
- 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.

Prevalence

- The 32nd Bethesda Conference estimated that the total number of adults living with congenital cardiovascular defects in the United States in 2000 was 800 000. In the United States, 1 in 150 adults is expected to have some form of congenital heart disease, including mild defects such as bicuspid aortic valve and more severe disease.
- The most common types of defects in children are ventricular septal defect (VSD), 620 000 people; atrial septal defect (ASD), 235 000 people; valvar pulmonary stenosis, 185 000 people; and patent ductus arteriosus, 173 000 people. The most common lesions seen in adults are ASD and tetralogy of Fallot (TOF).

Unless otherwise noted, all statistics in this Fact Sheet pertain to the United States.

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Mortality

- Mortality related to congenital cardiovascular defects in 2018 was 2,903 for all ages.
- In 2018, the age-adjusted death rate attributable to congenital cardiovascular defects was 0.9 death per 100,00 people, an 18.2% decrease from 2008.
- In 2018, congenital cardiovascular defects were the most common causes of infant death resulting from birth defects; 21.8% of infants who died in 2018 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 the air pollutant benzene 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.
- 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.
- Preeclampsia is a risk factor for congenital heart defects, although not critical defects.
- 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 in one study, 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).
- In 2016, 45,000 U.S. adults and children (25,000 males; 20,000 females) diagnosed with congenital heart defects were discharged from hospitals.

Congenital Cardiovascular Defects - 2021 Statistical Fact Sheet

For additional information, charts and tables, see
[Heart Disease & Stroke Statistics – 2021 Update](#)

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

Many statistics in this Fact Sheet 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 factsheet, 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:

Virani SS, Alonso A, Aparicio HJ, Benjamin EJ, Bittencourt MS, Callaway CW, Carson AP, Chamberlain AM, Cheng S, Delling FN, Elkind MSV, Evenson KR, Ferguson JF, Gupta DK, Khan SS, Kissela BM, Knutson KL, Lee CD, Lewis TT, Liu J, Loop MS, Lutsey PL, Ma J, Mackey J, Martin SS, Matchar DB, Mussolino ME, Navaneethan SD, Perak AM, Roth GA, Samad Z, Satou GM, Schroeder EB, Shah SH, Shay CM, Stokes A, VanWagner LB, Wang N-Y, Tsao CW; on behalf of the American Heart Association Council on Epidemiology and Prevention Statistics Committee and Stroke Statistics Subcommittee. Heart disease and stroke statistics—2021 update: a report from the American Heart Association [published online ahead of print January 27, 2021]. *Circulation*. doi: 10.1161/CIR.0000000000000950

If you have questions about statistics or any points made in the 2021 Statistical Update, please contact the American Heart Association National Center, Office of Science & Medicine at statistics@heart.org. Please direct all media inquiries to News Media Relations at <http://newsroom.heart.org/newsmedia/contacts>.