American Heart Association.

2019 Heart Disease & Stroke Statistical Update Fact Sheet
Congenital Cardiovascular Defects

CCDs arise from abnormal formation of the heart or major blood vessels. CCDs range in severity from very minor abnormalities that will never require medical therapy or intervention to complex malformations, including absent or atretic portions of the heart, that could require multiple surgeries and interventions, or even 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 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%
- Coarcation 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

- In 2002, it was estimated that there were 650,000 to 1.3 million children and adults living with congenital cardiovascular defects in the United States.
- In the United States, 1 in 150 adults is expected to have some form of congenital heart disease, including mild defects such as a well-functioning bicuspid aortic valve and more severe disease.
- The most common types of defects in children were ventricular septal defects (VSD), 93,000 children; atrial septal defect (ASD), 78,000 children; valvular pulmonary stenosis, 58,000 children; and patent ductus arteriosus, 58,000 children. The most common lesions seen in adults were ASD, 109,000 people and VSD, 106,000. This is based on 2002 data for children and adults and excludes bicuspid aortic valve.

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

Mortality

- Mortality related to congenital cardiovascular defects in 2016 was 3,063 for all ages.
- In 2016, the age-adjusted death rate attributable to congenital cardiovascular defects was 1.0 death per 100,00 people, a 16.7% decrease from 2006.
- In 2016, congenital cardiovascular defects were the most common causes of infant death resulting from birth defects; 22.0% of infants who died in 2016 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 mellitus (DM), including gestational DM, has also been associated with cardiac defects, both isolated and multiple. Pre-gestational DM is also associated with congenital heart defects, specifically tetralogy of Fallot (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 tetralogy of Fallot (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).
- The cost of identifying a newborn with critical congenital heart defects has been estimated at $20,862 per newborn detected and $40,385 per life-year gained (2011 US dollars).
- In 2014, 39,000 U.S. adults and children (21,000 males; 18,000 females) diagnosed with congenital heart defects were discharged from hospitals.

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

For additional information, charts and tables, see
Heart Disease & Stroke Statistics – 2019 Update

Additional charts may be downloaded directly from
https://www.ahajournals.org/doi/10.1161/CIR.0000000000000659 or
https://www.heart.org/en/about-us/heart-and-stroke-association-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:
Benjamin EJ, Muntner P, Alonso A, Bittencourt MS, Callaway CW, Carson AP, Chamberlain AM, Chang AR, Cheng S, Das SR, Delling FN, Djousse L,Elkind MSV, Ferguson JF, Fornage M, Jordan LC, Khan SS, Kissela BM, Knutson KL, Kwan TW, Lackland DT, Lewis TT, Lichtman JH, Longenecker CT, Loop MS, Lutsey PL, Martin SS, Matsushita K, Moran AE, Mussolino ME, O'Flaherty M, Pandey A, Perak AM, Rosamond WD, Roth GA, Sampson UKA, Satou GM, Schroeder EB, Shah SH, Spartano NL, Stokes A, Tirschwell DL, Tsao CW, Turakhia MP, VanWagner LB, Wilkins JT, Wong SS, Virani SS; on behalf of the American Heart Association Council on Epidemiology and Prevention Statistics Committee and Stroke Statistics Subcommittee. Heart disease and stroke statistics - 2019 update: a report from the American Heart Association [published online ahead of print January 31, 2019]. *Circulation.* doi: 10.1161/CIR.0000000000000659.

If you have questions about statistics or any points made in the 2019 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.

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