

Statistical Fact Sheet 2018 Update

Congenital Cardiovascular Defects

Congenital cardiovascular defects are structural problems that arise from abnormal formation of the heart or major blood vessels. ICD-9 lists 25 congenital heart defect codes, of which 21 designate specific anatomic or hemodynamic lesions; however, there are many more lesions that are not well described by ICD-9 or ICD-10 codes because of the wide diversity of congenital cardiovascular malformations, as well as varying classifications of nomenclature. Defects range in severity from tiny septal communications or defects between chambers that can resolve spontaneously to major malformations that may require multiple surgical procedures in infancy. In fact, on occasion a lesion can be so severe that it leads to in utero demise. As such, congenital heart defects are serious and common conditions that have significant impact on morbidity, mortality, and healthcare costs in children and in 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

- The most commonly reported incidence of congenital heart defects in the United States is between 4 and 10 per 1,000, clustering around 8 per 1,000 live births.
- Continental variations in birth prevalence have been reported, from 6.9 per 1000 births in Europe to 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.

Congenital Heart Defects - 2018 Statistical Fact Sheet

Prevalence

- In 2002, it was estimated that there were an estimated 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 are expected to have some form of congenital heart disease, including mild defects such as a well-functioning bicuspid aortic valve as well as 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 2002 data for children and adults and excludes bicuspid aortic valve.

Mortality

- Mortality related to congenital cardiovascular defects in 2015 was 3,128. .
- The age-adjusted death rate attributable to congenital cardiovascular defects was 1.0 death per 100,00 people, a 16.7% decrease from 2005.
- Congenital cardiovascular defects are the most common cause of infant death resulting from birth defects; 24.0% of infants who died in 2015 of a birth defect had a heart defect.
- In studies looking at trends since 1979, age-adjusted death rates declined 22% for critical congenital heart defects, 39% for all congenital heart defects, and deaths tended to occur at progressively older ages.

Risk Factors

- Numerous intrinsic and extrinsic nongenetic risk factors contribute to 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.
- 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-accepted risk for congenital defects, including congenital heart defects, and folic acid supplementation is recommended during pregnancy.
- Paternal exposures that increase risk for congenital heart defects include paternal anesthesia, sympathomimetic medication, pesticides, solvents and in one study, phthalates.

Congenital Heart Defects - 2018 Statistical Fact Sheet

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.

For additional information, charts and tables, see
[Heart Disease & Stroke Statistics – 2018 Update](#)

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

The American Heart Association requests that this document be cited as follows:
Benjamin EJ, Virani SS, Callaway CW, Chang AR, Cheng S, Chiuve SE, Cushman M, Delling FN, Deo R, de Ferranti SD, Ferguson JF, Fornage M, Gillespie C, Isasi CR, Jimenez MC, Jordan LC, Judd SE, Lackland D, Lichtman JH, Lisabeth L, Liu S, Longenecker CT, Lutsey PL, Matchar DB, Matsushita K, Mussolino ME, Nasir K, O'Flaherty M, Palaniappan LP, Pandey DK, Reeves MJ, Ritchey MD, Rodriguez CJ, Roth GA, Rosamond WD, Sampson UKA, Satou GM, Shah SH, Spartano NL, Tirschwell DL, Tsao CW, Voeks JH, Willey JZ, Wilkins JT, Wu JHY, Alger HM, Wong SS, Muntner P; on behalf of the American Heart Association Council on Epidemiology and Prevention Statistics Committee and Stroke Statistics Subcommittee. Heart disease and stroke statistics—2018 update: a report from the American Heart Association [published online ahead of print January 31, 2018]. *Circulation*. DOI: 10.1161/CIR.0000000000000558.

If you have questions about statistics or any points made in the 2018 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>.