Congenital Cardiovascular Defects

Congenital cardiovascular defects, also known as congenital heart defects, are structural problems that arise from abnormal formation of the heart or major blood vessels. ICD-9 lists 25 congenital heart defects codes, of which 21 designate specified anatomic or hemodynamic lesions. Defects range in severity from tiny pinholes between chambers that may resolve spontaneously to major malformations that can require multiple surgical procedures before school age and may result in death in utero, in infancy, or in childhood.

The common complex defects include the following:
- Tetralogy of Fallot (TOF)
- Transposition of the great arteries
- Atrioventricular septal defects (ASD)
- Coarctation of the aorta
- Hypoplastic left heart syndrome

Incidence
- Congenital heart defects are serious and common conditions that have significant impact on morbidity, mortality, and healthcare costs in children and adults.
- 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 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
- It is estimated that there are 850,000 adults and 859,000 children living with congenital heart disease in the United States in 2000.
- In the United States, 1 in 150 adults are expected to have some form of congenital heart disease.
- The most common types of defects in children are (at a minimum) ventricular septal defects, 620,000 people; ASD, 235,000 people; valvular pulmonary stenosis, 185,000 people; and patent ductus arteriosus, 173,000 people. The most common lesions seen in adults are ASD and TOF.

Mortality
- Mortality related to congenital cardiovascular defects in 2011 was 3,166. Any-mention mortality related to congenital cardiovascular defects in 2011 was 4,900.
- In 2011, there were 1,342 deaths due to congenital heart defects in white males, 1,117 in white females, 291 in black males, and 258 in black females.
- Congenital cardiovascular defects are the most common cause of infant death resulting from birth defects; 24% of infants who die of a birth defect have a heart defect.
- In studies looking at trends since 1979, age-adjusted death rates declined 22%37 for critical congenital heart defects, 39% for all congenital heart defects.
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.
- A greater risk of congenital heart defects is also seen in women who both have a high BMI.
- Gestational DM has also been associated with cardiac defects, both isolated and multiple.
- Folate deficiency is a well-accepted risk for congenital defects, including congenital heart defects, and folic acid supplementation is recommended during pregnancy.

Hospitalizations & Costs
- In 2004, hospital costs for congenital cardiovascular defect conditions totaled $2.6 billion. The highest aggregate costs were for stays related to cardiac and circulatory congenital anomalies, which accounted for ≈$1.4 billion, more than half of all hospital costs for birth defects.
- 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 2010, 62,000 U.S. adults and children (38,000 males; 24,000 females) diagnosed with congenital heart defects were discharged from short-stay hospitals.