Congenital Heart Defects

KNOW THE FACTS

Congenital Heart
PUBLIC HEALTH CONSORTIUM

What is a congenital heart defect?

Congenital heart defects (CHDs) are problems present at birth that affect the structure or function of the heart. They can affect how blood flows through the heart and out to the rest of the body.

There are many types of heart defects, with different degrees of severity based on size, location, and other associated defects. Common examples include holes in different areas of the heart and narrow or leaky valves. In more severe forms of CHDs, blood vessels or heart chambers may be missing, poorly formed, or in the wrong place.

What is the difference between congenital heart defect and congenital heart disease?

These terms are often used interchangeably and are nearly synonymous. However, there is a slight difference between them. A congenital heart defect refers specifically to a problem with the formation of the structure of the heart or major heart vessels in utero. Congenital heart disease refers to the clinical manifestation of an underlying anatomical defect, or more broadly describes functional problems which may be congenital, including certain arrhythmias.

CHDs include holes in the inside walls of the heart and narrowed or leaky valves. In severe forms of CHDs, blood vessels or heart chambers may be missing, poorly formed and/or in the wrong places.

What are critical congenital heart defects?

Critical CHDs (CCHDs) are defects that typically result in low oxygen levels in the newborn. Babies with critical CHDs usually require surgery or other procedures in the first year of life. Some examples of critical CHDs include coarctation of the aorta, transposition of the great arteries, hypoplastic left heart syndrome, and tetralogy of Fallot.[1]

Detection

When can CHDs be detected?

CHDs can be detected as early as the prenatal period or as late as adulthood (or escape detection altogether).[2] The more severe the form of CHDs, the more likely it is to be detected earlier.[3]

Approximately **120 infant deaths are prevented** each year with pulse oximetry screening

How can CHDs be detected?

There are a number of tools that can be used to aid in the diagnosis of CHDs, including echocardiogram, electrocardiogram, chest X-ray, chest CT, cardiac MRI, and prenatal ultrasound.[4] One or more of these diagnostic tests may be ordered if a healthcare provider finds a reason to suspect that the child has a CHD or if the child fails a newborn screening test.

What is newborn screening for critical congenital heart disease?

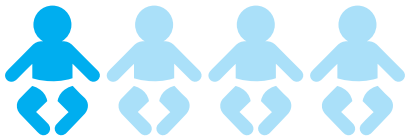
Newborn screening for critical congenital heart disease uses pulse oximetry to check the level of oxygen in the blood of newborns. This test is painless and non-invasive, which means that nothing is inserted into the newborn. In 2011, critical congenital heart disease was added to the United States Recommended Uniform Screening Panel. It is estimated to prevent approximately 120 infant deaths each year from critical congenital heart disease.[5, 6] As of 2018, policies for screening newborns have been implemented in all states in the U.S. Children who fail this screening can then have further testing to evaluate for congenital heart disease or for other potential causes of low oxygen.[1]

Prevalence

How many babies are born each year with some form of CHD?

Of the nearly 4 million infants born in the United States each year, approximately 3% have some type of birth defect.[7] CHDs are the most common birth defect, occurring in about 1 in 110 births, or nearly 1% of births. Thus, in the U.S. approximately 40,000 infants are affected each year.[3, 9-11]

Worldwide birth prevalence has been relatively stable since 1995 but has significant geographic variation. On average, about 1 in 110 births are affected, representing approximately 1.35 million live births with CHD each year.[8]



CHDs occur in nearly 1% (1 in 110) of births and **about a quarter are life-threatening.**

How many babies are born each year with a critical CHD?

Approximately 1 in every 4 babies born with a CHD has a critical CHD and needs surgery or other procedures in the first year of life.[9, 10]

How does the prevalence of CHDs at birth compare to other childhood and adult disease states and defects?

For comparative purposes, cystic fibrosis occurs in 1 in 3,400 live births,[11] autism is diagnosed in 1 in 59 children aged 8 years,[12] and new diagnoses of cancer are made in 1 in 6,024 children and teens annually.[13] Thus, the public health significance of CHDs in terms of numbers of people affected is much greater than or similar to that of disorders having higher levels of public awareness.

What is the most common form of CHDs in children?

In children, the most common form of CHDs is the ventricular septal defect, a hole between both main lower chambers, or ventricles, of the heart. The severity of the defect depends on its size and other associated anomalies.[3, 9, 16]

How many children and adults with CHDs are living in the U.S.?

Because there is no population-based surveillance of CHDs across the lifespan in the United States, little data are available on children, adolescents, and adults living with CHDs. Based on Canadian data from 2010[14] extrapolated to U.S. Census data, it is estimated that approximately 1 million children (966,000-993,000) and 1.4 million adults (1.41 – 1.46 million) were living in the U.S. in 2010, of whom approximately 12% (290,000) had severe CHDs. These estimates are increasing over time, especially for adults, as children with more severe defects are having a greater likelihood to survive to adulthood.

How does the prevalence of CHDs vary around the world?

Worldwide birth prevalence of CHD has been relatively stable. Asia has the highest reported birth prevalence of 9.3 per 1,000 (1 in 108 births) and Africa the lowest of 1.9 per 1,000 (1 in 526 births).[8] The highest reported total CHD birth prevalence was in high-income countries.[8] Because many of these births occur in developing countries that lack adequate care for CHD, hundreds of thousands of children die each year from CHDs. In high-income countries, over 85% of children with CHDs survive into adulthood, and as such, the worldwide burden of adult CHDs is expected to increase.[15] In most Western countries, the number of adults with CHDs outnumbers the children with an increasing margin. In Canada from 2000-2010 the prevalence of CHDs rose nearly six times faster among adults than children, and by 2010 adults accounted for nearly 66% of the CHD population.[14]

Mortality

How do CHDs affect the infant mortality rate in the U.S.?

All birth defects combined account for approximately 20% of infant deaths.[16] CHDs are the most common cause of infant death related to birth defects, accounting for 23.5% of infant deaths due to birth defects in 2014.[17]

How many deaths occur from unrecognized CHD?

The use of variable definitions and terminology make it difficult to accurately determine the number of deaths from unrecognized CHD. Furthermore, with improved prenatal detection of CHD and newborn screening for

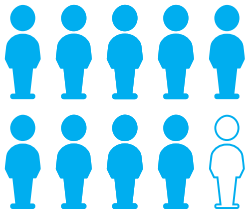
critical CHD, there is expected to be a decrease in the number of infants with unrecognized CHD. Prior to the advent of newborn screening for critical CHD, it was estimated that at least 280 infants with unrecognized critical CHD were discharged from the newborn nurseries each year in the United States.[18] Based on results from a study that examined the association of having a state policy for critical CHD screening and infant cardiac death, it is estimated that approximately 120 lives are saved each year via newborn screening for critical CHD.[6]

How many people in the U.S. die each year from CHDs?

From 1999 to 2006 there were 19.4 million deaths in the United States. Of these, 41,494, or 0.21%, were related to CHDs.[19]

How is a person's lifespan affected by CHD?

With improved treatment, people with CHDs are living longer, with over 85% of children born with CHDs living into adulthood. Following guidelines for coordinated and continued care has been shown to decrease mortality and thus may improve lifespan, health, and quality of life.[20, 21] However, studies have shown that individuals with CHDs often fall out of appropriate care. For example, 47-60% of young adults in Canada aged 18-22 never received recommended follow-up care.[22] In 2005, among all those with CHDs who were less than 55 years of age living in the United States, 192,000 total years were not lived in good health because of their condition. This loss in years of good health is approximately comparable to the many years of good health lost due to leukemia, prostate cancer, and Alzheimer's disease combined.[17, 23]



85-90% of patients born with severe CHDs who undergo surgery are expected to survive to age 18

What percentage of people with CHDs undergoing surgery survive the surgery?

In 2017, surgical survival (i.e. those without operative mortality) for congenital heart disease surgery in the Society of Thoracic Surgeons database was 97.3% overall. By age group this was 92.6% for newborns (<1 month old), 97.4% for infants (1 month to 1 year of age), 98.9% for children ages 1-18 years, and 98.7% for adults.[24] Operative mortality includes (1) all deaths occurring during the hospitalization in which the operation was performed, even if after 30 days; and (2) all deaths occurring after discharge from the hospital, but before the end of the thirtieth postoperative day.[25] From 1988 to 2003, the in-hospital death rate for adults undergoing surgery for CHD was shown to be significantly lower when the surgery was performed by pediatric vs. non-pediatric heart surgeons.[26]

How has mortality due to CHDs changed over time?

Mortality associated with several types of CHDs has decreased among both children[27] and adults,[28] with more people living to adulthood and thereby increasing the prevalence of CHD among adults.[29] Survival among infants with critical CHD improved from 67% in the era from 1979-1993 to 82.5% in the era from 1994-2005.[10] In the United States, among those who were operated on between 1982 and 2003 who survived their first CHD surgery, mortality declined over time, but it remains higher (8.3 times) than the general population and varies by severity of disease (35 times greater for single ventricle CHD; 4.3 times greater for mild forms of CHD).[30] In Sweden, survival improved for individuals with CHD who were born more recently, but there was less improvement in survival among adults with CHD.[31, 32]

What percent of individuals receiving care for severe CHDs are still alive at age 18?

In the current era, 85-90% of patients born with severe CHDs who undergo surgery are expected to survive to age 18.[9, 29, 33] This varies widely by type of defect, however. Survival for severe two-ventricle lesions such as transposition of the great arteries is close to 98% in the current era.[34] On the other hand, for the most severe defects such as hypoplastic left heart syndrome, survival to even 5 years can be as low as 38%.[35] However, much of the mortality is in the first year. Among those with hypoplastic left heart syndrome who survive to one year of age, survival to adolescence and adulthood approaches 90%.[36]

Are there racial and ethnic differences in death rates due to CHDs?

Death rates due to CHDs have been found to be lower among Hispanics (0.7/100,000 females, 0.8/100,000 males) than whites (0.8/100,000 females, 1.1/100,000 males) and blacks (1.1/100,000 females, 1.3/100,000

males).[37] The risk of in-hospital mortality has been noted to be higher among Hispanics (1.22) and blacks (1.27) compared with whites.[37]

Risk factors and causes

What factors can lead to CHDs?

Although some CHDs have been linked to genetic disorders, maternal conditions, and environmental factors, the causes for the majority of CHDs are unknown.[38, 39]

Most causes of CHDs are unknown. **Only 15-20% of all CHDs are related to known genetic conditions.** Expecting parents should talk to their doctor about their health and family history.

What genetic factors have been linked to CHDs?

At least 15% to 20% of CHDs have been linked with known genetic disorders. Of these, the more commonly seen are Down syndrome and other types of trisomies, Turner syndrome, and 22q11.2 deletion. With the use of available new technology, the ability to identify genetic disorders that are related to CHDs will increase.[40, 41]

What non-genetic factors have been linked to CHDs?

While the causes of most CHDs are unknown, research has shown that women who are obese, have pre-existing diabetes, use tobacco and/or take certain medications during pregnancy may have an increased risk of having a baby with a CHD.[38, 39] Other studies have indicated that parental age, use of assisted reproductive techniques, prior pregnancies, and maternal chronic conditions such as epilepsy, and use of medications, such as valproate, phenytoin, and isotretinoin may be associated with the occurrence of some types of CHDs.[42] Finally, psychosocial factors including socioeconomic status and stress have been shown to be associated with CHD risk.[39]

Despite finding factors that can increase the risk of having a baby with a CHD, evidence showing that modifying these risk factors would decrease the occurrence of CHDs are inconclusive. Furthermore, some studies may indicate risks potentially associated with particular CHDs but not with other CHDs. One study estimated that the proportion of identifiable, modifiable risk factors may be as high as 30% for some defects.[43] While minimizing risk factors does not eliminate the possibility of having a child with a CHD, reducing risk factors before and during the first trimester of pregnancy when the heart is forming may help prevent CHDs.

What can I do to minimize the chance that my child will have a CHD?

Prospective mothers should discuss any medical conditions and health behaviors that may affect a pregnancy, such as nutrition, physical activity, lifestyle, and occupation with their health care providers.

In particular, women of childbearing age should take multivitamins containing folic acid daily both before and during pregnancy, avoid tobacco and alcohol use, use only medications necessary for maternal health, and achieve a healthy weight before pregnancy. Women with diabetes should be in good glycemic control before becoming pregnant.[38, 44] Planning for pregnancy may help avoid inadvertent harmful exposures to the fetus in the first trimester, when mothers may not yet realize they are pregnant.

In addition, women of childbearing age should obtain preconception and prenatal care, including testing for diabetes and past rubella exposure. They should discuss any medication use with their physicians, and avoid contact with anyone who is ill, especially with febrile respiratory illnesses.[38, 45]

Do most people with CHDs have other family members with these conditions?

Some persons with CHD do have other family members with similar conditions. This familial association is more common with parents and siblings than with other relatives. In a study of sibling risks of CHD, CHD in a firstborn child is associated with increased risk for CHD in subsequent children.[46] The types of CHDs that may occur among family members may be different, however. The majority of persons with CHDs have no other family members exhibiting CHD.[40] Reasons for familial links in CHD conditions could be a combination of shared genes and environmental factors.[46]

Are people with a CHD more likely to have children with CHDs? How much more likely?

Parents with CHDs are more likely to have children with CHDs than those without CHD. An infant born to a family without a close relative with CHD has about a 1% chance of having CHD. The risk increases about three-fold for a family in which the mother, father, or sibling has CHD.[40]

Morbidity and other non-medical considerations over the lifespan

Do people with CHDs have other genetic or congenital health problems?

About 20% to 30% of persons with CHDs have other birth defects or genetic disorders, and may have developmental and cognitive disorders. Such physical abnormalities and developmental disorders may differ among those with different types or even the same type of CHD.[47-50]

About 50% of children with CHD may require special education services. About 50% of youth and adults with CHD experience anxiety or depression.

How many surgeries are performed on children and adults each year to treat CHDs?

According to the Society of Thoracic Surgeons Congenital Heart Surgery Database, in 2017 there were 30,172 surgeries performed on 24,996 patients. Of these, 51% were newborns or infants, 40% were between 1 and 18 years of age, and 10% were adults.[24]

How many surgeries does an adult with a CHD have during his or her lifetime?

Many people with CHDs are not cured with one surgery. They may develop problems that need additional surgeries or interventions. The need for more surgery depends on the type of CHD, initial operation type, and the year it was performed.[51-54] In one specialized cardiac center, almost 50% of adults with CHD who had surgery for their condition had two or more operations.[55]

Are other procedures besides surgery available for children and adults with CHDs?

There are catheter-based procedures for a variety of interventions, such as closure of a secundum atrial septal defect, occlusion of a patent ductus arteriosus, or placement of a pulmonary valve.[56] The choice of whether to do a catheter-based intervention or surgery depends on a number of factors, including the technical difficulty of the procedure, the nature of the defect, and patient preference.

What acquired cardiac conditions are of concern for individuals with CHD?

In addition to managing the CHD, affected individuals remain at risk for developing acquired cardiac conditions of adulthood including both those related and unrelated to the CHD.[57-60] These include:

- Arrhythmias[61-65]
- Atherosclerotic heart disease[66]
- Congestive heart failure[67]
- Endocarditis[68]
- Hypertension[66, 69-71]
- Hyperlipidemia[66]
- Pulmonary hypertension[72]

The risk of these conditions will vary by type of CHD and prior procedures.

Pregnancy can be high risk for women with congenital heart defects. It is crucial that women with CHD obtain care from a team of experts during pregnancy.

What do women with CHD need to know about reproductive health?

CHD is now the most common form of heart disease during pregnancy in the United States.[60] Because pregnancy among women with CHD may increase the risk of maternal and infant morbidity and mortality, guidelines have been developed for the management of pregnancy and preconception care.[102] Women with CHD should discuss their health concerns with an Adult Congenital Heart Disease (ACHD) cardiologist before becoming pregnant to determine how the pregnancy may affect them or the future baby.[20]

Multiple types of birth control are available for women with CHD. Each type has risk and benefit that vary with the underlying heart condition or type of repair.[20] For example, estrogen-containing birth control can increase the risk of clot formation and is not appropriate for patients with an underlying risk of clotting such as those with mechanical valves or who have had a Fontan operation.

If a woman with CHD is pregnant, it is recommended that she have an evaluation by a specialized multi-disciplinary care team at the time of pregnancy to formulate a care plan.[20, 102-104] As previously stated, CHD occurs in about 1% of births, with increased risk to 3-8% depending on parental CHD. Thus, fetal echocardiography is recommended at approximately 18-20 weeks gestation to screen for a CHD in the fetus.[105-107]

What are important neurodevelopmental, neurocognitive, and psychosocial considerations for individuals with CHD?

CHD is known to have important effects on the brain and brain development, with important ramifications both during childhood and adulthood.[108] People with CHD have a spectrum of such outcomes: many may notice few to no neurodevelopmental or neurocognitive defects, while others may have severe difficulties in life. During childhood, there can be challenges in neurodevelopment and academic achievement in school.[109-111] Into adulthood, there are various psychosocial considerations such as employment, social relationships, or mental health challenges.[112] It is therefore important that individuals with CHD, particularly those with the most severe forms, be screened for neurodevelopmental and neurocognitive challenges and be connected with services that may be of assistance.[113] With awareness, early identification and management of the neurodevelopmental, neurocognitive, and psychosocial challenges, the quality of life as perceived by those with CHD can be quite good.[114, 115]

What other acquired non-cardiac conditions are of concern for individuals with CHD?

Individuals with CHD are at risk of non-cardiac conditions that may be related to the underlying heart condition or may be sequelae of prior procedures or ongoing physiology. The prevalence of these conditions will differ depending on the underlying CHD and prior procedures. As some of these conditions may have a significant impact on quality of life and/or mortality, it is important for healthcare providers to be aware of these complications related to different CHDs or procedures and to assess patients appropriately.[73] Table 1 lists some of these potential non-cardiac conditions.

Table 1

System	Condition
Pulmonary	Restrictive lung disease[74-77] Plastic bronchitis[78] Chylothorax[79]
Renal	Chronic renal insufficiency[80]
Gastrointestinal/ Nutrition	Obesity[81-84] Malnutrition[84, 85] Protein losing Enteropathy[86] Hepatitis C[87, 88] Fontan-associated Liver Disease[89, 90] Chronic congestive hepatopathy[91, 92]
Neurologic	Seizure disorder[93] Cerebrovascular accident (stroke)[94, 95] Dementia[96]
Hematologic	Iron Deficiency anemia[97] Secondary erythrocytosis[97, 98]
Oncology	Potential increased risk of malignancy[99-101]

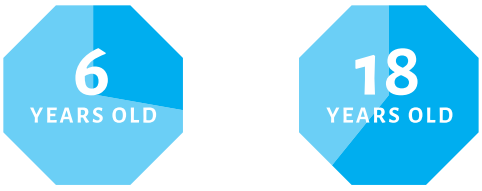
What mental health challenges are often encountered by individuals with CHD?

Although mental health challenges certainly do not affect every person with CHD, it has been shown that adults with CHD have a higher incidence of mental health issues than other heart healthy adults. Estimates range from approximately 36% of adults with CHD experiencing a diagnosable psychiatric disorder, with anxiety or depressive symptoms being prominent,[116] to at least 50% of interviewed patients meeting diagnostic criteria for at least one lifetime mood or anxiety disorder.[117] Unfortunately, most adults with CHD "suffer silently and worry alone", and many are not referred to a mental health professional.[118] According to one study, "approximately, 70% of the patients who met diagnostic criteria at the time of study participation were not engaged in mental health treatment."[117]

Healthcare Access and Cost Considerations

What is the national cost burden for treating congenital heart disease?

Cardiac and circulatory anomalies accounted for more than one-third of all hospital stays for birth defects and had the highest in-hospital mortality rate.[119] In 2013, hospital costs for all individuals with CHD exceeded $6.1 billion; this represents 27% of all birth defect-associated hospitalization costs.[120] Costs vary by age and type of CHD. Costs for infants less than 1 year of age, for hospitalizations for critical CHDs, and for hospitalizations in which the patient died were highest. For example, among privately insured children in 2005 in the United States, it was estimated that the average medical cost for children with CHD younger than 3 years old was 10 to 20 times greater than for children of the same age without CHD.[121] The greatest cost difference was in the care of infants less than 1 year old.



28% of children with a CHD will stop seeing a cardiologist by the time they are 6 years old, increasing to 61% by age 18 years. Don't be a lost-to-care statistic.

In 2009, estimates from a nationwide database showed the hospital cost for 27,000 children treated primarily for CHD was nearly $1.5 billion and for about 12,000 adults it was at least $280 million.[122] However, these are minimum cost estimates for all those with CHD, as they do not reflect the total reimbursement hospitals actually received and do not include costs for: discharges where CHD was not the primary condition, inpatient doctor care, medications, outpatient care, or nonmedical costs to families such as transportation, lost wages, or child care.

In the US, admissions to the hospital via the emergency department increase during transition years from pediatric to adult care settings.[123] All adults with CHD, even those with mild to moderate disease, have significantly higher rates of health care utilization than their age-matched peers. Data from Canada have shown that adults with severe CHD use three to four times more health services and that those with other forms of CHD use twice as many health services than the general population.[124]

Does early detection play a role in reducing healthcare costs for critical congenital heart disease?

From 1998 to 2007, approximately 23% of infants with critical congenital heart disease were diagnosed after birth hospitalization.[125] Late detection (detection after birth hospitalization) was associated with 52% more hospital admissions, 18% more hospitalized days, and 35% higher inpatient costs during infancy. Infants whose CCHD is detected at birth hospitals experience cost savings in terms of hospitalization. Potentially preventable deaths occurred in 1.8% of infants with late detection of CCHD.[125] These findings support the recommendation for increased CCHD screening.

What are important health insurance considerations for those with CHD?

Lack of health insurance has been shown to be associated with increased mortality among infants with complex congenital heart disease.[126] Moreover, lack of insurance can lead to significant financial burdens for families affected by congenital heart disease.[127]

Adults with CHD may encounter significant challenges in obtaining or utilizing health insurance.[128] It is important not only for individuals with CHD to have appropriate health insurance coverage, but also that such coverage includes the necessary access and benefits unique to this special population.[129]

What percentage of adults with CHD receive care at a specialty adult congenital heart defect program?

With approximately 1.4 million adults with CHD in 2010[19] yet fewer than 100,000 visits to adult CHD centers in 2016[130], it is estimated that fewer than 10% of adults who might benefit from adult CHD programs are in such programs. Since 2015 the American Board of Internal Medicine has been certifying ACHD physicians to fill a critical need for specialized healthcare for the growing population of adults with CHD. However, as of 2018, ACHD centers are only available in 42 states and

Washington DC, and in more than half of these states there is only one specialty clinic for the entire state.

What are reasons persons with CHD are lost to care?

Although most individuals with CHD require life-long cardiology care, many adults are lost to cardiac follow-up or may experience gaps in care up to 10 years. Current guidelines recommend a guided transition be initiated in early adolescence to encourage continuity of care with adult providers. Despite this recommendation, lapses in care are common. This population is vulnerable to loss to follow-up from cardiology care: about 28% do not see a CHD provider after age 6, 47% after age 13, and 61% after age 18.[22] Approximately 42% of adults with CHD presenting at an adult CHD clinic had a gap in care of greater than 3 years. Some of the leading reasons for gaps in care include changes or loss of insurance, financial barriers, feeling healthy, the inaccurate belief that their defect has "been cured," failure to track appointment timing, and decreased parental involvement. Most losses to follow-up or gaps in care begin during the late teen years, during the transition from pediatric to adult care.[131]

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