

CONGENITAL CARDIOVASCULAR DISEASE

What is congenital cardiovascular disease or congenital heart disease?

Congenital (kon-JEN'ih-tal) means inborn or existing at birth. Among the terms you may hear are congenital heart defect, congenital heart disease and congenital cardiovascular disease. The word "defect" is more accurate than "disease." A congenital cardiovascular defect occurs when the heart or blood vessels near the heart don't develop normally before birth.

What causes congenital cardiovascular defects?

Congenital cardiovascular defects are present in about 1 percent of live births. They're the most common congenital malformations in newborns. In most cases scientists don't know why they occur. Sometimes a viral infection causes serious problems. German measles (rubella) is an example. If a woman contracts German measles while pregnant, it can interfere with how her baby's heart develops or produce other malformations. Other viral diseases also may cause congenital defects.

Heredity sometimes plays a role in congenital cardiovascular defects. More than one child in a family may have a congenital cardiovascular defect, but this rarely occurs. Certain conditions affecting multiple organs, such as Down's syndrome, can involve the heart, too. Some prescription drugs and over-the-counter medicines, as well as alcohol and "street" drugs, may increase the risk of having a baby with a heart defect. Researchers are studying other factors.

What are the types of congenital defects?

Most heart defects either obstruct blood flow in the heart or vessels near it, or cause blood to flow through the heart in an abnormal pattern. Rarely defects occur in which only one ventricle (single ventricle) is present, or both the pulmonary artery and aorta arise from the same ventricle (double outlet ventricle). A third rare defect occurs when the right or left side of the heart is incompletely formed -- hypoplastic heart.

This defect patent ductus arteriosus allows blood to mix between the pulmonary artery and the aorta. Before birth there's an open passageway (the ductus arteriosus) between these two blood vessels. Normally this closes within a few hours of birth. When this doesn't happen, some blood that should flow through the aorta and on to nourish the body returns to the lungs. A ductus that doesn't close is quite common in premature infants but rather rare in full-term babies. If the ductus arteriosus is large, a child may tire quickly, grow slowly, catch pneumonia easily and breathe rapidly. In some children symptoms may not occur until after the first weeks or months of life. If the ductus arteriosus is small, the child seems well. If surgery is needed, the surgeon can close the ductus arteriosus by tying it, without opening the heart. If there's no other defect, this restores the circulation to normal.

Obstruction defects

An obstruction is a narrowing that partly or completely blocks the flow of blood. Obstructions called **stenoses** can occur in heart valves, arteries or veins.

The three most common forms are pulmonary stenosis, aortic stenosis and coarctation of the aorta. Related but less common forms include bicuspid aortic valve, subaortic stenosis and Ebstein's anomaly.

Septal defects

Some congenital cardiovascular defects let blood flow between the heart's right and left chambers. This happens when a baby is born with an opening between the wall (**septum**) that separates the right and left sides of the heart. This defect is sometimes called "a hole in the heart."

The two most common types of this defect are atrial septal defect and ventricular septal defect. Two variations are Eisenmenger's complex and atrioventricular canal defect.

Cyanotic defects

Another type of heart defect is congenital cyanotic (kon-JEN'ih-tal si"ah-NOT'ik) heart defects. In these defects, blood pumped to the body contains less oxygen than normal. This causes a condition called cyanosis (si"ah-NO'sis), a blue discoloration of the skin. The term "blue babies" is often applied to infants with cyanosis.

Examples of cyanotic defects are tetralogy of Fallot, transposition of the great arteries, tricuspid atresia, pulmonary atresia, truncus arteriosus and total anomalous pulmonary venous connection.

Hypoplastic left heart syndrome

In hypoplastic (hi"po-PLAS'tik) left heart syndrome, the left side of the heart is underdeveloped -- including the aorta, aortic valve, left ventricle and mitral valve. Blood returning from the lungs must flow through an opening in the wall between the atria, called an atrial septal defect. The right ventricle pumps the blood into the pulmonary artery, and blood reaches the aorta through a patent ductus arteriosus. (See above.)

The baby often seems normal at birth, but will come to medical attention within a few days as the ductus closes. Babies with this syndrome become ashen, have rapid and difficult breathing and have difficulty feeding. This heart defect is usually fatal within the first days or months of life without treatment.

This defect isn't correctable, but some babies can be treated with a series of operations or with a heart transplant. Until an operation is performed, the ductus is kept open by intravenous (IV) medication. Because these operations are complex and different for each patient, you need to discuss all the medical and surgical options with your child's doctor. Your doctor will help you decide which is best for your baby.

Some doctors will recommend a heart transplant to treat this problem. Although it does provide the infant with a heart that has normal structure, the infant will require lifelong medications to prevent rejection. Many other problems related to transplants can develop. You should discuss these with your doctor.

Children with hypoplastic left heart syndrome require lifelong follow-up by a cardiologist for repeated checks of how their heart is working. Virtually all the children will require heart medicines. **People with hypoplastic left heart syndrome, before and after treatment, are at risk for getting an infection on the heart's inner lining or valves (endocarditis). To help prevent this, they'll need to take antibiotics before certain dental and surgical procedures.** Good dental hygiene also lowers the risk of endocarditis. For more information about dental hygiene and preventing endocarditis, ask your cardiologist.