What is congenital heart disease (CHD)? CHD is a term used to describe a group of problems present at birth that affect the structure of the heart and/or the large vessels near the heart. Congenital [kuhn-JEN-i-tl] means a problem present at birth. There are many types of CHD. Some types may be detected before birth and, in some cases, may be treated before birth. Other types are not found until the baby is born.

The effects of CHD range from mild to severe and depend on the type of heart defect and on the child’s general health. At one time, most infants born with CHD did not survive. Today, there are new treatments for CHD that are helping many children survive and lead a healthy life.

What causes CHD? Some forms of heart problems are passed through families (called genetic conditions). Other types of CHD are caused by problems that affect the fetus while in the womb. These factors may include problems in the mother, such as a disease, infection, or a substance to which either the mother or father has been exposed (such as alcohol or some types of medicines or solvents). These factors often have their greatest effect on the fetus during the first 2 to 7 weeks of pregnancy.

A brief list of some types of CHD. The picture on the left shows the outside of a normal heart. The numbers show you the general areas where the following types of CHD may occur.

1. **Atrial septal defect (ASD)** — at birth, there is a natural opening in this area that closes within several days. If it does not close on its own, ASD occurs.
2. **Ventricular septal defect (VSD)** — a hole that occurs in this area.
3. **Tetralogy of Fallot** — a group of defects that occur at the same time. This may include VSD, problems with the pulmonary valve and some associated problems.
4. **Coarctation of the aorta** — one part of the aorta is very narrow, so blood cannot flow through well.
5. **Transposition of the great vessels** — the great vessels (the aorta and the pulmonary artery) are reversed (they develop on the side of the heart opposite from where they should be placed).
6. **Truncus arteriosus** — the aorta and pulmonary arteries are joined as one common vessel.
7. **Tricuspid atresia** — the tricuspid valve does not open or is not present.
8. **Total anomalous pulmonary venous return (TAPVR)** — the pulmonary veins enter the wrong side of the heart.
9. **Hypoplastic left heart syndrome (HLHS)** — the left side of the heart is not developed.

The type of CHD my child has is called:
What happens in CHD? CHD affects the heart in one or both of the following ways:

Causing a detour in blood flow: some defects make the blood flow through the heart in an abnormal way and may then cause one part of the heart to work too hard, leading to heart pump failure and other problems. A defect in the septum, the wall that separates the left side of the heart from the right side, often leads to this problem. This type of problem also may occur if one part of the heart is not fully formed at birth (called hypoplasia [hie-puh-PLEY-zhuh]).

Blocking blood flow: some defects block blood flow to the heart and/or to the body. The blockage may occur in the heart or in the large vessels near the heart.

How do we know if our baby has CHD? If doctors suspect a baby has a high risk for CHD, they may look for signs of a defect while the baby is still in the womb. This involves giving the mother a test (called a fetal echocardiogram), which provides a picture of the baby’s heart structure and function. This test allows doctors to see most types of heart defects. The test may be done on the baby after birth if doctors suspect the baby has CHD. The test is similar to an ultrasound and poses no risk to either the mother or baby.

After birth, babies with CHD may show some of these signs:
- poor muscle tone (limp muscles)
- heartbeat is too fast (called tachycardia [tak-i-KAHR-dee-uh])
- breathing problems
- feeding problems
- overall very pale color
- heartbeat is too slow (called bradycardia [brad-i-KAHR-dee-uh])
- heart murmur — a whooshing sound that occurs when blood does not flow through the heart in a normal manner
- heartbeat is uneven (slow at times, fast at others)
- blue color to the hands, arms, legs, and feet

Doctors may use a pulse oximeter [ok-SIM-i-ter] to test the amount of oxygen in the baby’s blood to see if the baby’s heart is working well. This test helps find some types of CHD that may not be found with other tests until days or weeks later.

What can be done for CHD? Some types of CHD improve without treatment. Other types may not affect the child’s health and do not need treatment. Diseases that require treatment may need either medicine(s) and/or surgery. For some types of CHD, you will be shown special ways to help your child with activity and movement (or exercise, as your child grows older), with feeding, and other care tasks that may be needed after surgery.

Medicines: Doctors may prescribe different medicines, depending on the nature of the heart defect. Medicines called diuretics [dahy-uh-RET-iks] help remove excess fluid from the body. Since one of the heart’s functions is to pump fluid through the body, these medicines help reduce the heart’s workload. Other medicines, such as digoxin
[dij-OK-sin], help improve the heart’s pumping action. Some medicines may be used to treat blockages in the great vessels. For more details about the medicines your child needs, speak with your baby’s doctors and nurses.

**Surgery:** Doctors may recommend surgery to repair holes or other defects of the heart and help restore blood flow. Sometimes the surgical repairs must be made in stages so the child may need several surgeries. The surgery is performed by a pediatric cardiac surgeon, a doctor with special training to perform heart surgery in infants and children. For some types of CHD, surgery may be performed using a device inserted through a small incision. Other types of repairs require a more invasive procedure. Some infants may require a pacemaker (a device that helps regulate the heartbeat) or open heart surgery.

After surgery, the child will need help with breathing and may use a ventilator. Sometimes the child may need an extracorporeal [ek-struh-kawr-PAWR-ee-uhl] membrane oxygenation (ECMO) machine to help with breathing and heart function. The ECMO machine takes over the work of the heart and lungs for a brief time so these organs can rest and heal. This equipment may be used until the child’s body is strong enough to carry out these functions on his or her own.

**What are the long-term effects of CHD?** Long-term effects vary for every child and for every type of CHD. One of the main effects that may occur in some children is injury to the brain and nervous system (the network of nerves that runs throughout the body, connecting the brain with all parts of the body). This problem may occur when the brain does not receive enough blood flow and oxygen because the heart is not working well. The degree of injury to the brain and nerves may range from mild to severe, and depends largely on a complex mixture of factors, such as:

- the length of time the child’s brain and organs have not had enough oxygen.
- limited movement. Without enough oxygen, babies become tired and cannot interact well with others around them. This event disrupts their ability to explore their environment, which then disrupts the active growth and development of the brain and nervous system that usually occurs between birth and one year of age.
- limited interaction with others. Parents who are concerned about their child’s health may try to protect the child from injury and illness. In some cases, this reaction can limit the child’s growth and learning because the child has fewer chances to interact with other adults and children.
- long stays in the hospital for surgery and/or other problems from CHD may limit social interaction and motor development that normally would occur.
- emotional detachment from parents due to feeding problems and/or other issues resulting from CHD.

These factors may result in problems with learning, language, balance and coordination, vision, and fine motor skills (such as using fingers and hands for tasks). Problems with behavior, balancing emotions, poor attention span, and interacting with others also may occur, but these problems can be subtle and may not be noticed until the child reaches school age.* In many cases, these problems are mild and may be reduced if they are addressed early on. A key step to help prevent such problems or to avoid long-term effects from them is taking your child to all follow-up checkups advised by your doctor. You may be advised to take your infant for checkups with a doctor who has special training in this area (such as a developmental pediatrician and/or a neurodevelopmental specialist).

What can parents do? Today more infants than ever before are able to live a healthy life with CHD. Yet, having a child with a heart defect still may pose a great challenge for both child and parents. Sometimes the challenges may stretch beyond infancy, into childhood. Below are a few useful tips to help yourself and your child along the road ahead.

Keep a balance: Often it can be hard for parents to balance their desire to protect their child’s health, while allowing the child to lead a normal life — in short, to be the same as children who do not have an illness. Some parents may protect the child too much due to fears for the child’s health. Others may not protect the child enough, in an effort to help the child feel normal. Studies of parents of children with CHD show that parents often have trouble setting limits and instilling discipline for this reason.* One way to help maintain a balance is to talk to your child’s doctor about your concerns.

Keep informed: Your child’s needs will change as your child grows. Some children will require treatment for CHD throughout life. A key task for parents is to remain aware of these changing needs and to find help when needed.

Be aware of your baby’s physical needs: Some children may require early help with motor skills. For example, feeding may be hard for infants with CHD because they are less able to breathe and suck from the bottle or breast at the same time. Since feeding time is a key time for bonding, problems with feeding can disrupt bonding. Talk to your baby’s doctors about where to find help with these issues.

Be aware of your child’s emotional needs: Having a severe illness that involves many types of treatment and time in the hospital can affect children’s emotions. Children do not yet know all the words or concepts to describe what they feel as a result of illness and treatment. This issue, along with any injury to the nervous system, may lead to problems with emotion and behavior in some children. There are many people who can help you and your child address these issues, such as a child life specialist, psychotherapist and/or a developmental pediatrician.


Find the research

NICHD Cochrane Neonatal Review Group
www.nichd.nih.gov/cochrane

Neonatology on the Web
www.neonatology.org

Find out more: these websites may be helpful

American Academy of Pediatrics
www.aap.org/parents

American Heart Association
www.americanheart.org

American Pediatric Surgical Association
www.eapsa.org/parents

Early Head Start National Resource Center
www.ehsnrc.org

Genetics Home Reference

Health on the Net Foundation
www.hon.ch

Healthy Steps for Young Children
www.healthysteps.org

Take a closer look at these Pediatrix topics

- pulse oximeter
- ventilator
- nervous system

Tools of the NICU
- A2

How babies’ senses develop
- B2

Topics A - F can be found online.

This information is for educational purposes only and is not intended to substitute for professional medical advice. Always consult with a health care professional if you have any questions about the health of your baby.