

ACHA Q and A: Congenitally Corrected Transposition of the Great Arteries (CCTGA)*

What is CCTGA?

Congenitally corrected transposition of the great arteries (CCTGA) is a rare heart defect in which the heart's lower half is reversed. It is also called L-TGA. It is different from and much less common than "regular" transposition of the great arteries (TGA or D-TGA). To understand CCTGA, it helps to first understand how a normal heart works.

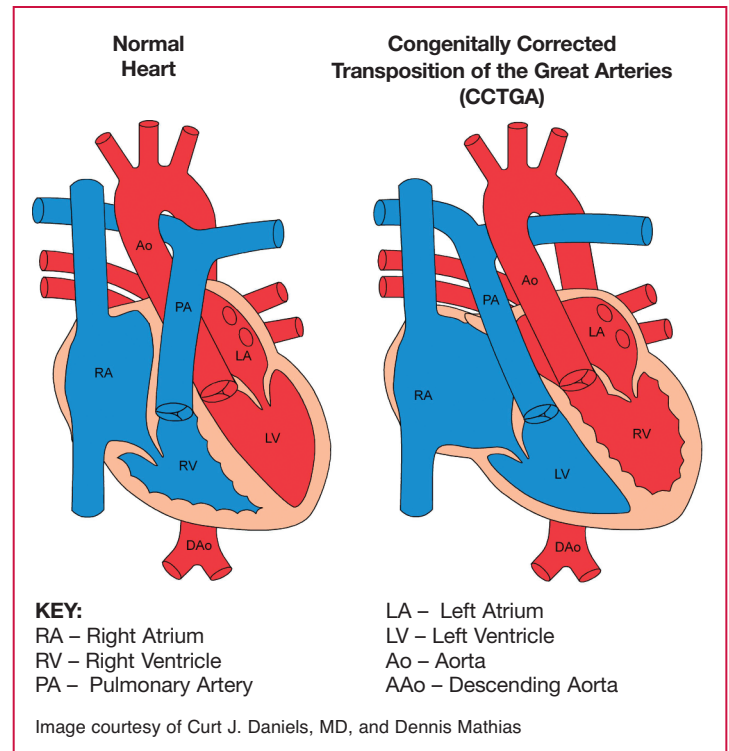
A normal heart is divided into two sides. The right side pumps blood from the body into the lungs. The left side pumps the blood from the lungs out to the body. Each side has an atrium and a ventricle. The atrium acts like a "waiting room" for the blood. The ventricle does the hard work of pushing the blood out to the lungs or body. At the entrance and exit from each ventricle is a valve, which acts like a door. These valves allow the ventricle to fill with blood from one side, and then push it out the other.

Each ventricle and valve is designed to do its specific job. The right ventricle is designed to give the blood a gentle push to send it to the lungs. It is bigger than the left ventricle and does not have as much muscle. The left ventricle is designed to give the blood a strong push out to the body. It has less space inside because its thick walls take up more space. The valve between the left atrium and ventricle, the mitral valve, is the body's strongest valve. It is designed to stay shut against the strong push of the blood out to the body.

About 0.5%–1% of all babies born with heart defects have CCTGA. This means there are about 5,000–10,000 people in the United States with CCTGA.

In CCTGA, the two ventricles and their attached valves are reversed. Your weaker, larger right ventricle grows on your heart's left side. It pumps your blood to your body. Your stronger, smaller left ventricle grows on your heart's right side. It pumps blood to the lungs. You also have your body's weakest valve—the tricuspid valve—serving as your mitral valve.

**Please note: This article is directed to those who have not undergone a double-switch repair.*



How common is CCTGA?

About 0.5%–1% of all babies born with heart defects have CCTGA. This means there are about 5,000–10,000 people in the United States with CCTGA.

How does CCTGA develop?

In the womb the heart starts out as a simple tube. It then bends in two, creating the right and left sides of the heart. In CCTGA, it folds in the wrong direction. The ventricle meant to grow on the right side is on the left. The ventricle meant to grow on the left is on the right. Because the valves are part of the ventricle, they are also reversed.

What causes CCTGA?

We do not know enough about what causes CCTGA. The studies we have now suggest that CCTGA is rarely passed from parent to child. One study reported that of 120 parents with CCTGA, none had babies with heart defects. Another 2001 study reported an increase of CCTGA in areas with certain toxins. More research is needed into the causes of CCTGA and other congenital heart defects.

How is CCTGA diagnosed?

Some babies born with CCTGA have heart murmurs, heart rhythm problems, and/or show signs of heart failure. If they

also have a ventricular septal defect (VSD) they may also be cyanotic (blue). In these cases a diagnosis is usually made in infancy. However, it is not unusual for CCTGA to be found later in childhood or adulthood. This is because you can have no symptoms and have CCTGA. Often it is the start of new heart symptoms that lead to a diagnosis of CCTGA. Sometimes it is found accidentally when routine heart tests are done.

Echocardiography can be used to diagnose CCTGA in adulthood. But the heart's unusual shape makes imaging CCTGA difficult. Many doctors now prefer cardiac MRI (magnetic resonance imaging) when evaluating CCTGA. For those with implanted devices that make MRI impossible, CT (computed tomography) scans can be used.

What is the long-term impact of CCTGA?

One of the mysteries of CCTGA is its wide range of outcomes. Some people born with CCTGA live into old age with no or few problems. On the other hand, some babies born with CCTGA die in infancy. Many people diagnosed with CCTGA have no or few symptoms as children and young adults, but develop more problems as they age. There are three common problem areas:

- **Heart rhythm problems**

Almost everyone born with CCTGA has an abnormal atrioventricular (AV) node. The AV node sends the electrical signals from the top to the bottom of the heart. Many people with CCTGA develop heart block. This means there is a blockage in the signal. Your heart may not be able to beat as quickly or efficiently as it should. In complete heart block, your atria and ventricles start to beat separately. This can cause light-headedness and fainting. In rare instances it can be fatal. Pacemaker therapy is used to treat heart block. Up to 45% of those with CCTGA need pacemakers.

- **Valve problems**

Many people with CCTGA have malformed tricuspid valves. The overload on this valve can also cause or worsen leaking. Replacement of this valve is sometimes recommended. When done early, replacing this valve can improve symptoms. Heart function may also be protected and/or restored. However, valve replacement can only be done if your heart function is still relatively good. Valve replacement in CCTGA should only be done at centers that specialize in ACHD surgery.

- **Heart failure problems**

Some people with CCTGA develop heart failure. Heart failure is when the heart is too weak to pump blood efficiently. Many medications now exist to help with heart failure. If heart function is only mildly weakened, valve replacement may also be considered. There are also new pacemaker therapies which can improve your heart's ability to serve your body. If heart failure becomes too severe, a heart transplant may be considered.

What other defects are common in CCTGA?

In addition to VSD and tricuspid valve problems (discussed earlier), pulmonary stenosis, single ventricle, and dextrocardia are also common. Pulmonary stenosis is a narrowing between the heart and the lungs. This can cause pressure problems in the heart. Up to 50% of those with CCTGA also have pulmonary stenosis. Single ventricle is when the hole in the heart is so big that the two ventricles merge. The right and left ventricles become two sides of a common chamber. About 40% of those with CCTGA also have a single ventricle. Dextrocardia is when the heart is on the right side of the chest. About 20% of people with CCTGA also have dextrocardia.

If you have CCTGA you should see your cardiologist regularly, even if you feel fine. New heart problems can occur with no symptoms.

Can women with CCTGA have children?

Most women with CCTGA can have children successfully. The exceptions are if your heart function is too weak, if you have serious lung problems, and/or if you have low oxygen levels in your body. If you have CCTGA, it is very important that you consult with a specialized ACHD clinic before you get pregnant. If you become pregnant, your ACHD cardiologist and your obstetrician will take special steps to protect your heart during your pregnancy and delivery.

What kind of cardiology care is recommended for adults with CCTGA?

The American Heart Association and the American College of Cardiology classify CCTGA as a highly complex heart defect. This means that experts recommend that you get your care at a special ACHD center. You can find a listing of ACHD clinics at www.achaheart.org.

If you have CCTGA you should see your cardiologist regularly, even if you feel fine. New heart problems can occur with no symptoms. Many treatments that can help strengthen and protect your heart work best if started early. It is also important that you continue to protect your heart from infection (endocarditis) by taking antibiotics as recommended by your cardiology team.

The good news is that most people with CCTGA continue to do well. One study showed that three out of four patients with isolated CCTGA were free from heart failure at age 45. By taking good care of your heart, and getting recommended care, you can help ensure that you and your "backwards heart" continue to thrive.

*Thank you to **Amy Verstappen** and **Drs. Karen Kuehl** and **Daniel Murphy, Jr.** for their contributions to this article. ■*