

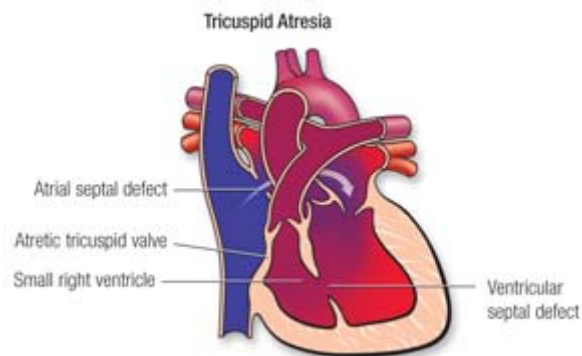
Single-Ventricle Defects

What is it?

Complex heart defects that result in one of the heart's pumping chambers (ventricles) being underdeveloped are called single-ventricle defects. Each of these defects is relatively rare. They include such problems as tricuspid valve atresia, hypoplastic left-heart syndrome, hypoplastic right-heart syndrome (pulmonary atresia with intact ventricular septum) and double-inlet ventricle. Other types of heart defects, such as atrioventricular canal defects or double-outlet right ventricle, may be complicated by an underdeveloped ventricle. The most common types of single ventricle defects are tricuspid atresia, pulmonary atresia/intact ventricular septum and hypoplastic left heart syndrome, and are described below. Read further to learn how they affect the heart, surgeries and ongoing care.

Tricuspid Atresia

In this condition, there's no tricuspid valve so blood can't flow normally from the right atrium to the right ventricle.



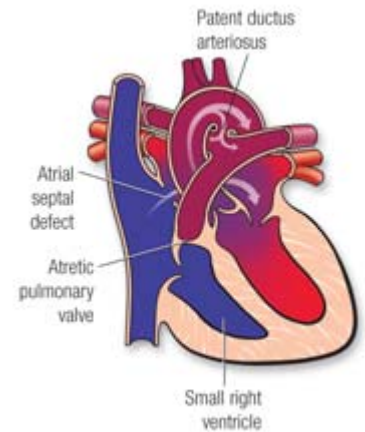
As a result, the right ventricle is small and not fully developed. Survival depends on there being an opening in the wall between the atria (atrial septal defect) and usually an opening in the wall between the two ventricles (ventricular septal defect). As a result, the low-oxygen (bluish) blood that returns from the body veins to the right atrium flows through the atrial septal defect and into the left atrium. There it mixes with oxygen-rich (red) blood from the lungs. Most of this partially oxygenated blood goes from the left ventricle into the aorta and on to the body. A smaller-than-normal amount flows through the ventricular septal defect into the small right ventricle, through the pulmonary artery, and back to the lungs. Because of this abnormal circulation, the patient with this condition looks blue (cyanotic) until surgery can be performed.

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Pulmonary Atresia/Intact Ventricular Septum

In pulmonary atresia, no pulmonary valve exists. Blood can't flow from the right ventricle into the pulmonary artery and on to the lungs. The right ventricle and tricuspid valve are often poorly developed.

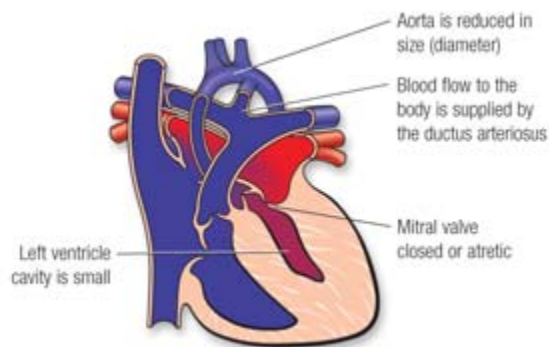
An opening in the atrial septum lets blood exit the right atrium, so low-oxygen (bluish) blood mixes with the oxygen-rich (red) blood in the left atrium. The left ventricle pumps this mixture of oxygen-poor blood into the aorta and out to the body. The infant appears blue (cyanotic) because there's less oxygen in the blood. The only source of lung blood flow is the patent ductus arteriosus (PDA), an open passageway between the pulmonary artery and the aorta.



Some patients with pulmonary atresia/intact septum can have a repair that allows the right ventricle to grow and function in a more normal way than other patients with single ventricles. A more complete repair depends on the size of the pulmonary artery and right ventricle. If the pulmonary artery and right ventricle are very small, the patient may require the same type of operation as other single ventricle patients. In some patients, abnormal channels (sinusoids) form between the coronary arteries and the right ventricle. These sinusoids can complicate this condition and even result in a heart transplant being recommended.

Hypoplastic Left Heart Syndrome

In hypoplastic left heart syndrome (HLHS), the heart's left side — including the aorta, aortic valve, left ventricle and mitral valve — is underdeveloped. Blood returning from the lungs must flow through an opening in the wall between the atria (atrial septal defect). The right ventricle pumps the blood into the pulmonary artery, and blood reaches the aorta through a patent ductus arteriosus.



The series of operations are similar to those done in other patients with single ventricles; however the first operation (Stage I Norwood) is more complicated than for other patients with single ventricles.

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What causes a single-ventricle heart defect?

In some cases, a genetic problem may be associated with a single-ventricle heart. Most cases have no identifiable cause.

How does it affect the heart?

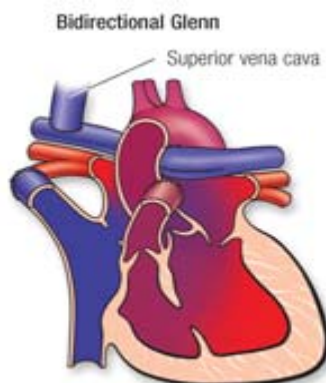
Underdevelopment of a ventricle can compromise blood flow to the body in some cases and the lungs in other cases. Without early intervention many patients die in infancy. In a small number of patients, the type of single ventricle results in a balanced circulation between the body and lungs, and survival through childhood is possible without surgery.

How does it affect me?

Almost all adults with single ventricles have had at least one, and in many cases, two or three operations in childhood. Most of these operations result in bluish (oxygen-depleted) blood from the veins going to the lungs without going through a pumping chamber (ventricle) and the ventricle that's present pumping blood to the body. Problems can arise from inefficiency of blood going to and returning from the lungs because of the lack of a pumping chamber to the lungs.

If single ventricle was repaired in childhood, what can I expect?

Surgery is often necessary in the first week of life to increase or decrease blood flow to the lungs. A procedure called a shunt is done to increase blood flow to the lungs in the first week of life. This improves the cyanosis. Some children with tricuspid atresia have too much blood flowing to the lungs. They may need a different type of surgery, called pulmonary artery banding, to decrease blood flow to the lungs. This is important to protect the lung blood vessels. A small number of children have just the right amount of blood going to the lungs and don't need an initial operation.

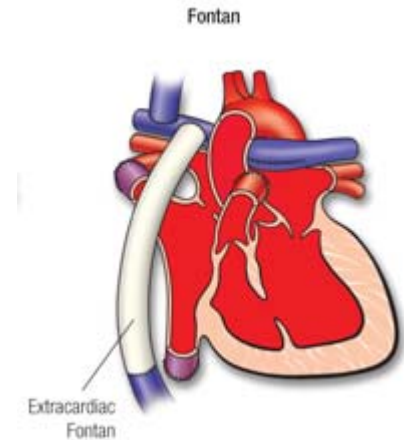


Almost all adults with single ventricle have also undergone subsequent operations to create connections between the body veins (inferior vena cava and superior vena cava) and the lung (pulmonary) arteries. This is usually done in two stages. First, the large vein from the upper half of the body (the superior vena cava) is connected to the lung arteries in a procedure called a bi-directional Glenn Operation.

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Later, the large vein from the lower half of the body (the inferior vena cava,) as well as the veins from the liver, are connected to the lung arteries in a surgery called a Fontan Operation.

Sometimes, at the time of the Fontan surgery, an opening, called a fenestration, is purposely left between the bluish (low-oxygen) and red (high-oxygen) sides of the blood flows. The Fontan operation may eliminate or greatly improve the cyanosis, but without a right ventricle that works normally, the heart doesn't work like a normal heart, which has two pumps.



Problems You May Have

Many patients have survived three to five decades after single ventricle surgery in childhood. Many of these patients are highly functional, with good activity levels. However, some patients with single-ventricle defects do have health problems.

In the few patients in whom surgery has not been performed, there is cyanosis (lower oxygen levels, causing blueness), lower energy and a higher risk of infections such as brain abscess or endocarditis (infection of the heart). These problems shorten the lives of some people.

If you've had surgery for a single-ventricle defect, you can live a relatively normal life. However, your ability to exercise vigorously is usually reduced.

Several basic types of problems are most common in this group of people. These problems may relate to the person's age at the time of the operation and the type of surgery done. Potential problems include:

1. Rhythm problems, generally fast heart rate (tachycardia, supraventricular tachycardia, atrial flutter) or slow heart rate.
2. Fluid retention, particularly in the abdomen and lower extremities. Some adults may develop varicose veins after the operation.
3. Greater risk of weakening and failing heart muscle when there's only one ventricle.
4. Blood clots inside the heart that may require anticoagulation therapy.

Ongoing Care

What will I need in the future?

Single-ventricle defects are among the most complex congenital heart problems. If you have this defect, you'll need regular checkups by cardiologists with expertise in adult congenital heart disease as well as ongoing care all your life. You should also consult a cardiologist with expertise in care of adult congenital heart disease if you're undergoing any type of non-heart surgery or invasive procedure.

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Medical

Many people with single-ventricle defects require daily or multiple medications. This care is best given by a cardiologist who's very familiar with the anatomical complexities and complications that these patients have. This requires the expertise of a cardiologist trained in congenital heart disease.

You will need at least yearly checkups to monitor your health. This may mean that you require such tests as an electrocardiogram (ECG), echocardiogram (ultrasound of the heart, including transesophageal echocardiograms), cardiac catheterization, Holter and arrhythmia event monitoring, and exercise stress-testing.

Activity Restrictions

You may need to limit your activity, particularly competitive sports. If you have decreased heart function or rhythm disturbances, you may need to limit your activity more. Your cardiologist will help you determine if you must limit your activities.

Endocarditis Prevention

Antibiotics to prevent endocarditis are needed before certain dental procedures.

Pregnancy

Some women who have had a Fontan-type operation can conceive and safely carry a pregnancy to term. The risk increases if the heart muscle is weak, if there is obstruction or clot in the Fontan connection or if there are arrhythmias. If you want to become pregnant, it's important to talk to your cardiologist before conception to find out the risks of pregnancy. It's also important to have a high-risk obstetrician who is experienced in caring for patients with congenital heart defects during pregnancy and delivery.

Will You Need More Surgery?

Most single ventricle surgeries are performed in the first two to four years of life and don't require more surgery. If you have any of the complications noted above, you may need to have prior surgical procedures revised. Additional interventions may include closing holes, pacemakers, or repair or replacement of leaky valves. In rare cases, a heart transplant may be considered. Your doctor will explain these possibilities, because each single ventricle has unique features.