

SINGLE VENTRICLE CONGENITAL HEART DEFECTS

What is a single ventricle heart defect?

A single ventricle congenital heart defect is the general name for many types of heart defects that result in having one functional ventricle, or pump. In some cases, one of the ventricles is hypoplastic, or small. In other situations, one of the valves that lets blood enter into the ventricles is missing (atresia) or does not let much blood go across it (stenosis). Some of the specific types of single ventricle congenital heart defects include: tricuspid atresia with ventricular septal defect, double inlet left ventricle, unbalanced atrioventricular septal defect, hypoplastic left heart syndrome. There are many other specific diagnoses that are classified as single ventricle congenital heart defects, but the main thing that brings them together is that instead of two ventricles, one to pump to the lungs and one to pump to the body, there is only one usable pump.

What causes these types of heart defects?

There is no known cause of these types of heart defects. Some patients may have other birth defects, and some have genetic disorders. Certain types of single ventricle heart defects are more likely to be associated with additional defects.

How are these defects diagnosed? What are signs and symptoms?

Single ventricle heart defects are often diagnosed by fetal ultrasound before the baby is born. First trimester screening for chromosomal abnormalities is a good screening tool to identify patients who might be at an increased risk for cardiac defects. Single ventricle heart defects can be diagnosed as early as 12 weeks gestation. In some cases, single ventricle heart defects develop during the pregnancy, and cannot be diagnosed until the second trimester targeted scan. Babies with single ventricle heart defects may initially appear healthy, although their oxygen levels will be slightly decreased. They may develop difficulty breathing, poor feeding, cool and pale extremities, or appear blue.

How will your pregnancy be managed?

If your baby is diagnosed with a single ventricle heart defect, a high-risk obstetrician will participate in your obstetric care. Overall care should be transferred to a specialized center, where multidisciplinary care is available. The team should include a perinatologist, fetal and pediatric cardiologists, a genetic counselor, a neonatologist and a pediatric cardiac surgeon. Fetal well being will be followed closely by fetal ultrasound and nonstress tests. Towards the end of pregnancy, visits may be as often as two to three times a week. If there is no specific maternal or fetal reason for a C-section, vaginal delivery is often possible. Induction of labor is often scheduled for pregnancies affected with single ventricle heart defects to make sure that all of the team members are available at the time of delivery.

Why do these heart defects make babies sick?

The left side of the heart is normally responsible for receiving blood rich in oxygen from the lungs and pumping that blood out to the body. The right side of the heart pumps oxygen-poor blood to the lungs to get oxygen. When there is one ventricle, it must do the work of both, pumping blood to the lungs and the body at the same time. The blood that the ventricle pumps is a mixture of oxygen-rich and oxygen-poor blood.

This is not a problem early on, because before birth and for a short time after birth, babies are able to use other routes to get blood to the body and the lungs. A connection exists between the pulmonary artery (artery from the right ventricle to the lungs) and the aorta (blood vessel from the left ventricle to the body). That connection, called a patent ductus arteriosus (PDA), is present in everyone before birth and closes within the first week of life.

Depending on the size and arrangement of the pulmonary artery and aorta, babies with single ventricle defects fit into a few main types of blood flow patterns. The pattern of blood flow determines what type of symptoms they have and how we will need to treat them in the first few weeks of life.

The first category is babies with not enough pulmonary blood flow, or blood flow to their lungs. These babies may have no pulmonary valve or a small pulmonary valve that limits the flow of blood to their lungs. These babies use the PDA to get extra blood to their lungs, but may become very blue if the PDA closes and little or no blood gets to their lungs.

Some types of heart defects result in not enough blood getting to the body. These babies have no aortic valve, a small aortic valve or a narrowing in the aorta, called a coarctation. The PDA will also help in this situation to provide blood flow to the body, but when it closes, the babies will have limited blood flow to the body. These babies present with rapid breathing, cool and pale extremities and can become quite ill.

In some cases, there is no limitation to blood flow to the lungs or body. These patients have normal sized pulmonary and aortic valves. Before birth, the pressure in the lungs is high, and there is less blood going to the lungs. This is because babies don't use their lungs before they are born. After birth, the pressure in the lungs drops. The pressure in the lungs becomes much lower than the pressure in the body, and thus it is easier for blood to flow to the lungs. Over time, there will be more blood going to the lungs than the body. The heart has to maintain a certain amount of blood to the body, but it may end up pumping 3-5 times more blood with each squeeze to compensate for the extra blood going to the lungs. When this happens, we say that there is too much pulmonary blood flow. These babies usually do very well right after birth, but within the first month of life may start to breathe fast or have difficulty feeding and gaining weight.

What can I expect after my baby is born?

Babies with these types of heart defects will need evaluation and stabilization in the neonatal intensive care unit after birth. The diagnosis will be confirmed with an echocardiogram, and babies may get large IVs placed. The immediate treatment for newborns with single ventricle defects depends on the blood flow patterns, as discussed above. If there is not enough pulmonary blood flow or not enough blood flow to the body, we can give a medication called prostaglandins to keep the PDA open. Babies with too much pulmonary blood flow typically don't need any treatment right away.

Because the care for children with congenital heart disease is complex, your baby will be cared for by a team of skilled clinicians. This starts before birth with maternal-fetal medicine and fetal cardiologists and continues with pediatric cardiologists and nurse practitioners, a pediatric cardiac surgeon, neonatal and pediatric intensive care physicians, pediatric anesthesiologists, pediatric cardiac operating room staff, pediatric nurses and many others.

What is the treatment/surgery for single ventricle defects?

Newborn surgery

These treatments help to stabilize the baby for surgery, but don't provide a long term solution. We don't yet have a way to make another ventricle, but there is a series of surgeries that allows us to use one ventricle as a pump and get blood to both the body and lungs. When there is not enough pulmonary blood flow, we can place a shunt, which is a small tube that connects the aorta to the pulmonary artery, which sends extra blood flow to the lungs. This acts just like a PDA. The surgery to place the shunt would be done in the first week of life. Babies usually stay in the hospital for 3-4 weeks after the surgery.

If the baby has not enough blood flow to the body, a procedure called the Norwood procedure may be needed. The Norwood operation is an open heart surgery that accomplishes 3 goals: getting blood to the lungs, getting blood to the body and making sure that the blood returning from the lungs can get to the right side of the heart. The surgeon attaches the pulmonary artery and the aorta to create one large blood vessel that goes from the right ventricle to the body. This means that the branches of the pulmonary artery that go out into the lungs are no longer attached to the heart. A tube is placed that connects either the right ventricle or the aorta to the pulmonary artery branches and creates a stable way to get blood to the lungs. Finally, the wall between the right and left atria is cut out to allow all the blood coming back from the lungs to get to the right side of the heart. This is performed in the first week of life, and babies stay in the hospital for about a month after surgery.

If there is too much pulmonary blood flow and babies develop symptoms which are not controlled with medications, then a surgery may be done to limit the blood flow to the lungs. A band is placed around the pulmonary artery to create a narrowing that will block some of the blood flow to the lungs. This might be done between 1-3 months of age, depending on how the baby is doing. Babies typically recover quickly from the procedure and are home within a week.

Some babies may be "well balanced", meaning that they have no limitation of blood flow to their bodies and just the right amount of blood flow to their lungs. These babies don't require an initial surgery in the first few months, and instead will go straight to the Glenn shunt (see below). These initial surgeries provide enough blood to both the lungs and the body, but they are not a final solution. The single ventricle has to do extra work, by pumping blood to both the lungs and the body, and the body gets blood that is a mixture of oxygen rich and oxygen poor blood. Also, a shunt (the tube that gets blood to the lungs) does not grow with the baby.

Later surgeries

When babies are 4 to 6 months of age, a second procedure is done. This is called a Glenn shunt, and it is the first step of separating the lung and body circulations. The large vein that drains the oxygen-poor blood from the head and upper body into the right atrium (called the superior vena cava, or SVC) is detached from the heart and attached directly to the pulmonary arteries. This lets the blood drain directly into the lungs. The tube connecting the right ventricle or aorta to the pulmonary artery is removed. After this procedure, the right ventricle only has to pump the blood to the body, so it doesn't have to work as hard. However, all of the oxygen-poor blood from the lower body is still coming into the right atrium by another large vein (called the inferior vena cava, or IVC) and mixing with the blood from the lungs. This means that the blood going to the body still has a decreased oxygen level. After this surgery, babies usually stay in the hospital for about 7-10 days.

The final procedure, called the Fontan operation, takes care of this problem and is usually done around 2 to 4 years of age. In this surgery, the IVC is removed from the heart and connected to the pulmonary arteries. At this point, all of the oxygen-poor blood will drain directly into the lungs, and the right ventricle will pump oxygen-rich blood to the body. After this surgery, children usually stay in the hospital for about 7-10 days.

These surgeries require a team of highly trained providers, like the team we have at the University of Maryland Children's Heart Program. Our team includes a pediatric cardiac surgeon, pediatric cardiologists, pediatric and neonatal intensive care doctors and nurses, pediatric cardiac anesthesiologists, cardiac operating room staff, cardiac nurses and many others.

What other procedures or follow up will my baby need?

Some patients, particularly those with a shunt, may need close monitoring between their initial surgery and the Glenn procedure. We instruct parents on what to look for and give you a machine to check their oxygen levels daily and a scale to weigh them daily. This allows us to pick up problems early. Some babies have difficulty feeding after the Norwood surgery and may not be able to take all their feeds by mouth. Some may require placement of a tube directly into their stomach to make it easier to get feeds.

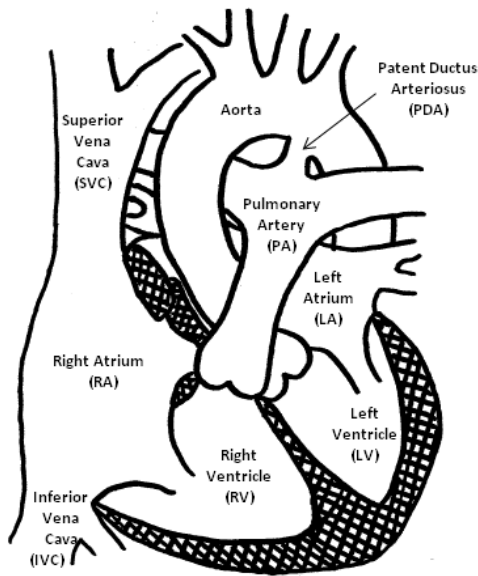
Children with single ventricle defects will need lifelong follow up with a cardiologist. In addition to the 3 surgeries that we expect, they may need other procedures. Most children undergo a cardiac catheterization procedure before the Glenn and Fontan surgeries. Although follow up visits are frequent when they are young, after completing the series of surgeries they will be seen every 6 to 12 months. At follow up visits, children will be checked regularly with ECGs and echocardiograms. Other tests, such as cardiac MRIs, exercise stress tests or Holter (24 hour ECG) monitors may be used.

What is the long term outcome for children with single ventricle heart defects?

Survival for infants with infants with single ventricle congenital heart defects has been improving over time. Surgeries performed just after birth require more recovery time, but survival is high. Because they have no pump to their lungs, it is harder for these patients to increase the output of their hearts. Therefore, patients may need to take medications and may have some limitations in their exercise ability. Despite this, they are typically able to have happy and productive lives.

If I have had one baby with a single ventricle heart defect, am I more likely to have other babies with a single ventricle heart defect?

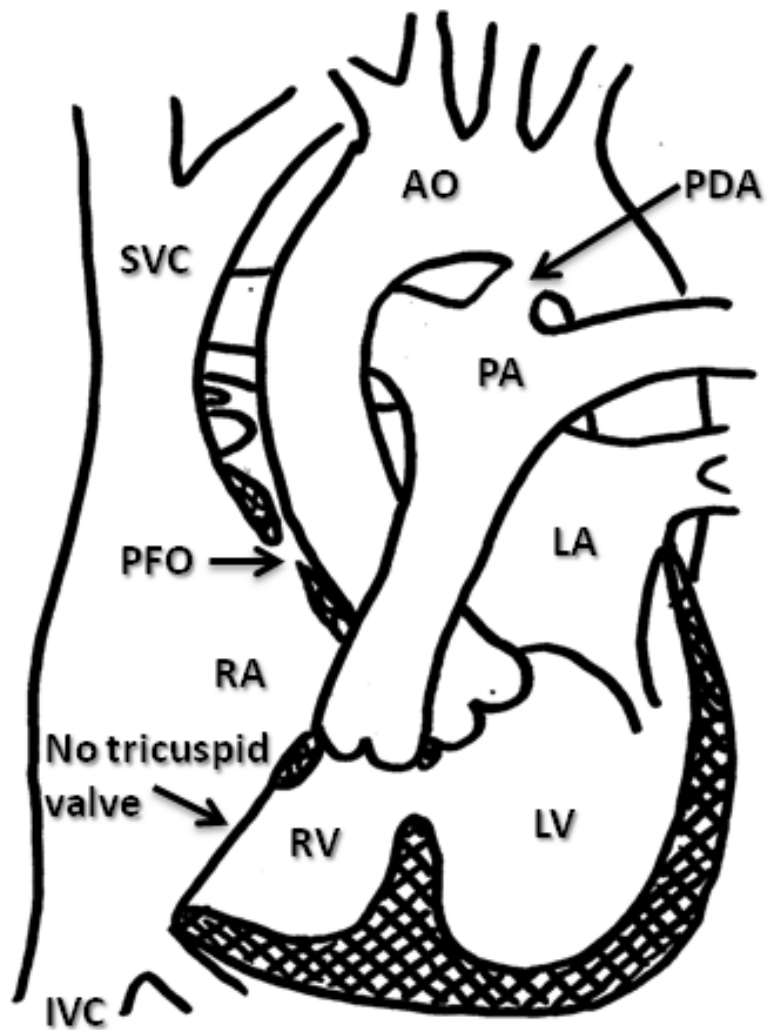
If your baby's single ventricle heart defect is related to a chromosome abnormality or a genetic syndrome, a genetic counselor can tell you what the chances are that a future pregnancy would have the same condition. When a single ventricle heart defect is not associated with an underlying genetic problem, future children are at a slightly increased risk for heart defects. A genetic counselor can tell you, based on the specific single ventricle heart defect your baby had, what the chances are that future children would have a heart defect. In future pregnancies, nuchal translucency ultrasound (at the end of the first trimester), targeted anatomy ultrasound (between 18-20 weeks) and fetal echocardiography are recommended.



Normal Newborn Heart

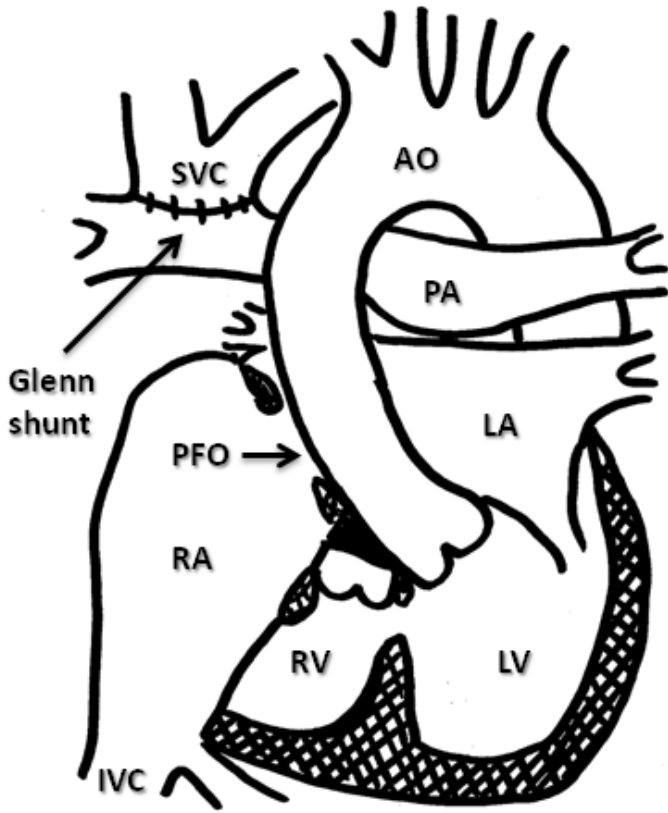
Tricuspid Atresia with Ventricular Septal Defect

(A type of single
ventricle defect)

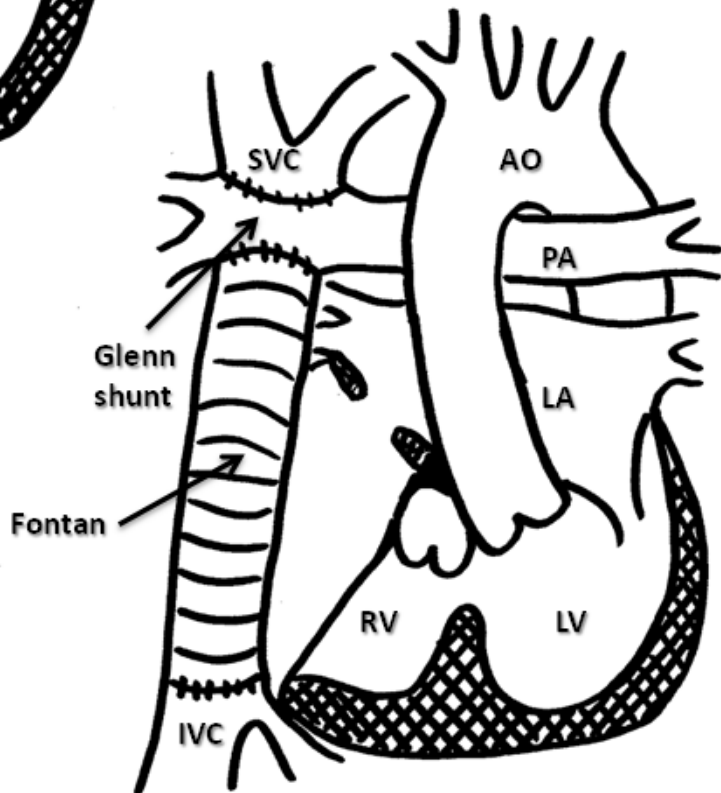
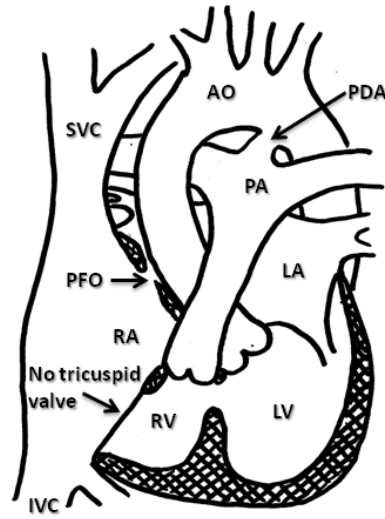


After
surgeries:

Before surgery:



Glenn



Fontan