

What is a “single ventricle”?

A single ventricle is a heart defect in which one lower chamber (ventricle) does not develop. This results in the heart having only one pumping chamber. Red blood (blood with oxygen) mixes with blue blood (blood without oxygen). The result is that the amount of oxygenated blood getting to the body is decreased. This causes cyanosis or blueness.

Single ventricle defects are rare. They occur in about five out of every 100,000 live births. Single ventricle defects include:

1. **Hypoplastic left heart syndrome (HLHS):** Occurs when the left ventricle, mitral valve, aortic valve and aorta are all undersized.
2. **Double outlet right ventricle:** Occurs when both the aorta and the pulmonary artery come out of the right ventricle. This leaves the left ventricle underdeveloped.
3. **Tricuspid atresia:** A defect where the tricuspid valve fails to develop. This leads to an underdeveloped right ventricle.
4. **Double inlet left ventricle:** occurs when both of the upper chambers of the heart (atria) connect to the left ventricle. This results in an underdeveloped right ventricle.

What causes single ventricles?

In most cases, the cause is unknown. In rare instances it may occur in families but no genetic link has been identified. Most often, single ventricle defects occur sporadically.

How are single ventricles treated?

In a normal heart, each ventricle does a separate job. The right ventricle pumps blood from the heart to the lungs. The left ventricle pumps blood to the body.

In a single ventricle heart, only one chamber is large enough to pump all of the blood. It is important that it is used efficiently and not overworked.

Surgical treatment for single ventricle starts early in the newborn period. It usually occurs in three stages. Each stage re-routes the blood. This allows the body to receive increasing amounts of oxygen. Doing it this way lessens stress on the heart due to the dramatic changes in the heart’s functionality with each procedure.

Depending on the degree of cyanosis and the amount of blood flowing to the lungs, either a shunt (Blalock-Taussig or Waterston shunt) or pulmonary artery banding procedure will be done in the first weeks of life. The shunt allows more blood to go to the lungs but there still continues to be mixing of red and blue blood. The pulmonary artery band prevents too much blood from circulating in the lungs.

The second procedure, the Glenn, replaces the first shunt in late infancy. Although this allows more oxygen-rich (red) blood to get

to the body, some mixing of blood still occurs. The final stage, known as the Fontan, is usually done when the child is between 1½ to 3 years old. The Glenn and Fontan ultimately separate the red and blue blood so that the child is no longer cyanotic.

When and where was the Fontan first done?

The Fontan procedure was originally performed by Dr. Francois Marie Fontan from Bordeaux, France in 1968 but not reported until in 1971. Over the years, modifications were made.

There have been three types of Fontans: the classic (or atrio-pulmonary), the lateral tunnel, or the extra-cardiac. The classic is not used anymore. It may be converted to one of the other types in an operation known as a Fontan revision.

How does a Fontan work?

In Fontan circulation, the way blood is pumped by the heart is changed. The single ventricle pumps blood returning from the lungs to the body. Blood returning is not pumped to the lungs. Instead, it flows passively from the body to the lungs by direct blood vessel connections from the lower and upper body to the lung arteries. Pressure changes caused by normal breathing and higher pressure in the veins help blood flow to the lungs. By allowing complete separation of blue blood and red blood, the Fontan may improve or get rid of cyanosis.

Are there any complications I need to watch for?

Adults with Fontan circulation should be followed regularly by an adult congenital heart disease (ACHD) specialist. Their anatomy puts them at risk for the development of some complications. These include arrhythmias, cyanosis, liver abnormalities, heart failure, protein losing enteropathy (PLE), blood clots, and pulmonary vascular disease (PVD).

Stage III – Fontan Operation

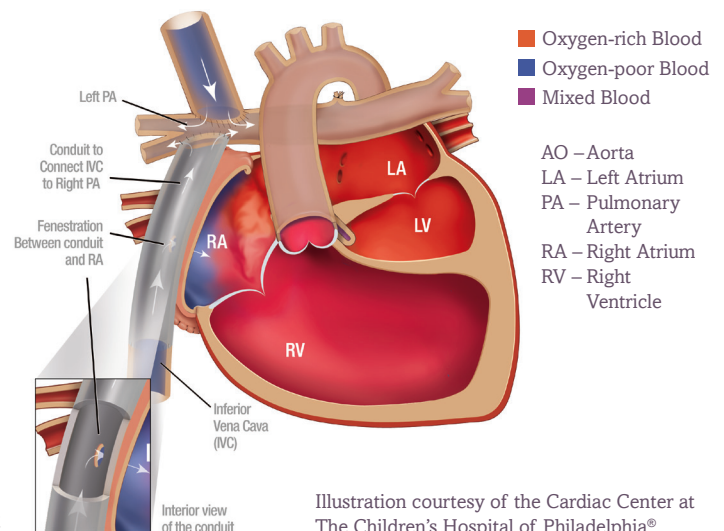


Illustration courtesy of the Cardiac Center at The Children’s Hospital of Philadelphia®

Arrhythmias

An arrhythmia is a problem with the rate or rhythm of the heart. It can be caused by scar tissue formation in the heart. In adults who have had the Fontan procedure, this presents as either a slow or fast heart rate. If the heart rate is too slow, a pacemaker might be necessary. While fast rhythms can happen in either the ventricle (lower chambers) or the atria (upper chambers), atrial flutter is the most common fast rhythm problem. Many times it can be treated with medication or ablation. Fast rhythms in the ventricle are rare but may require an automatic implantable cardioverter defibrillator (AICD).

Cyanosis

Cyanosis can develop after the Fontan. This can occur for a variety of reasons. These include leaks in the Fontan baffle or artificial wall, a fenestration (a surgically created opening in the Fontan circulation), an abnormal connection between arteries and veins in the lungs, or the development of extra blood vessels in the body, known as collaterals. Early treatment of these can prevent the development of more serious problems.

Liver complications

Blood returning to the heart in adults with Fontan circulation is at higher pressures than in hearts of people with two pumping chambers. This, along with decreased cardiac output, causes blood to flow back into the liver. This causes congestion in the liver. The liver in Fontan patients can also become thickened or fibrotic. Most adults with Fontan circulation have some degree of liver disease. Knowing who should be screened—as well as how and when to do so—is unclear.

Leg Varicosities

Adults with Fontan circulation have decreased blood flow to the legs and increased systemic pressures. As a result, chronic venous insufficiency is common and severe. Blood has trouble flowing back to the heart and pools in the legs. This can cause varicose veins and leg ulcers. Your ACHD doctor may recommend that you wear graduated compression knee highs with the greatest pressure at the foot and that you keep your ankles higher than your heart when sitting or lying down. Always check with him/her before doing this.

Protein Losing Enteropathy (PLE)

PLE is a rare (occurring in less than 10% of all Fontan patients) but serious complication. With PLE, the body does not absorb protein from the gastrointestinal tract and thus, it spills into the “belly.” This results in abdominal swelling, pain, blood clots, and diarrhea. Little is known about why it happens or how to treat it. We do know that it does not seem to occur as often in adults as it does in younger patients.

Heart Failure

Over time the heart of the adult with a single ventricle may become weak and not pump as well as it had been. This is known as heart failure. Heart failure can also occur because of leaky valves, valvular stenosis, obstruction in the Fontan, or narrowing in the lung arteries or veins. Heart failure can be treated with medications. It may also require further intervention by catheter or surgery.

Blood Clots

Blood clots may develop in people with Fontan circulation or in those with cyanosis. This increases their risk of having a stroke or pulmonary emboli. To prevent clots from forming, they may be placed on blood thinners such as warfarin or aspirin.

Pulmonary Vascular Disease

Blood flows passively to and from the lungs in Fontan circulation. The maintenance of low resistance is important to Fontan physiology. Pulmonary vascular disease or pulmonary hypertension is a possible long-term complication in Fontan patients.

Can I exercise?

Low to moderate level exercise is recommended in most patients with a Fontan. This includes walking, swimming and biking. Contact sports and heavy weightlifting should be avoided. It is best to check with your ACHD cardiologist prior to beginning any exercise program. Your ACHD team can advise you on what’s appropriate for you.

Can I get pregnant?

Women who have had a Fontan procedure may be able to have a successful pregnancy and delivery. However, the risk is high to both the mother and the baby. There is also a higher risk of obstetric complications, including miscarriage and preterm labor. Anyone with Fontan circulation should have a complete evaluation by an ACHD specialist and high-risk obstetrician before becoming pregnant and during the pregnancy.

The risk of passing on CHD to the baby is between 5% and 14% higher than the general population. This is true regardless of which parent has Fontan circulation.

What about birth control?

Women with Fontan circulation are at increased risk of blood clots. They should only use progesterone-based contraceptives. If sterilization is considered, it should be planned with the congenital cardiologist and a gynecologist familiar with Fontan circulation.

What if I need non-cardiac surgery?

Non-cardiac surgery in the patient with a Fontan should be done in an ACHD center. It should be closely coordinated with the ACHD team. An anesthesiologist with expertise in congenital cardiology is necessary.

What kind of cardiology care is recommended for adults with a Fontan procedure?

The American College of Cardiology and the American Heart Association classify all patients with Fontan circulation as having highly complex congenital heart disease. Experts recommend that you get care for your heart at a special ACHD center. You can find a listing of ACHD clinics at www.achaheart.org. If you have had a Fontan procedure, you should see your adult congenital cardiologist regularly, even if you feel fine. This way you can catch any problems before they start, since new heart problems can develop without symptoms. By taking good care of your heart, and getting recommended care, you can help ensure that you and your heart continue to thrive.