

## What is A Fontan Operation?

A Fontan operation refers to a type of operation that is used to help patients that have only one functioning Ventricle. It has helped to prolong the lives of many children who would have otherwise died in early adulthood. The most common of these are Tricuspid Atresia, Pulmonary atresia, Single Ventricles and Hypoplastic Left Ventricle. Normally the human heart has two ventricles. The right ventricle pumps blood from the veins that drain the body and sends it to the lungs where it picks up oxygen again. This oxygenated blood is then returned to the left side of the heart to the left atrium and from there to the left ventricle. The left ventricle is the main pump of the heart, which sends blood to the different parts of the body (head, arms, legs, kidneys, etc)

With the Fontan operation a patient only has one ventricle that must be used to pump blood to the various body parts, In this situation the blood is literally sucked through the lungs by the pumping action of the one ventricle, there is also some help with the movement of breathing and the diaphragm. Many variations of the Fontan operation exist as it has been modified significantly over the past 25 years. Originally it was designed to help children with Tricuspid atresia a condition where the tricuspid valve doesn't open the right ventricle is too small and the right atrium is large) It was originally thought that it was best to maintain the pumping action of an even small right ventricle to move blood to the lungs. Valves use to be placed in the right atrium and pulmonary artery in order to simulate a more "normal " situation. As the years went by it has become apparent that the least resistance to flow is the best way to create a Fontan circulation. For this reason we no longer put valves on the pulmonary side of the Fontan circulation. Most people are now using a staged approach to the Fontan. In this case the superior vena cava (vein that drains the upper part of the body) is attached to both pulmonary arteries at about 3-6 months of age. This is called a bi-directional Glenn shunt. At about a year or so later the inferior vena cava (drains the lower part of the body) is attached to the lungs as well. The method of how this attachment is done has been controversial over the past years and most recently the "extra-cardiac' Fontan has been used the most. This is where is plastic tube is used to extend the inferior vena cava to the pulmonary artery. Sometimes this cannot be done because the child is too small. The plastic will not grow. Also it is very common to create a small opening between the systemic and pulmonary veins or connection called a **fenestration**. This allows blood to flow directly to the ventricle if the pressure in the systemic veins is too high. This allows the patient to have an adequate cardiac output at the price of having a less than normal amount of oxygen in the blood.

The Fontan operation is not a cure for one ventricle hearts, and has associated with it many problems. These problems can be broken down into four parts . 1- relatively low cardiac output. Very frequently the valve associated with the systemic ventricle will leak and on occasion needs to be repaired or replaced. 2- higher venous pressure for the systemic veins. Because the blood is passively pushed through the lungs the pressure in the systemic veins (blood going to the lungs) is higher than in the veins coming back from the lungs. In order for there to be forward movement of blood through the lungs the pressure on the systemic side has to be higher than the pulmonary venous side. This creates a problem with the little blood vessels and capillaries in the body. On occasion the capillaries will begin to leak since they are subjected to higher pressures than normal and so this can cause a loss of protein and can cause a condition called Protein Losing Enteropathy or PLE this can be difficult to treat and can cause a buildup of fluid in the body (lungs, abdomen) and can cause bad diarrhea as well as loss of important minerals. 3- risk of irregular heartbeats or arrhythmias ). When the pressure in the right atrium is increase it causes the wall to stretch and the right atrium becomes enlarged. If it is stretched too much this can cause the heart to have irregular heart beats that can be very dangerous ( Atrial flutter and fibrillation). Sometimes medications and pacemakers can help control this. 4- Development of venous bypasses around the lungs. Because there is a pressure difference within the lungs that is opposite of normal over time small connections are developed that directly connect the systemic veins to the pulmonary veins. These occur mostly in the lungs, but can occur at other places as well. This causes the patient to become progressively bluer with time (normally after a Fontan a patient should have normal oxygen saturation) because blood that should go to the lungs for oxygen is bypassing the lungs and not being refilled with oxygen. Sometimes these extra connections can be coiled with a catheter but they tend to come back. Also there is a sense that because of abnormal flow patterns within the lungs over time there is a progressive blockage of the very tiny blood vessels within the lungs and this causes over time a rise in the pressure in the lungs. This in turn causes a progressive rise in the pressure in the systemic veins and can make conditions such as PLE worse and also hasten the development of those extra connections between the systemic and pulmonary veins.

If you have any questions, please ask one of the doctors.