

What is Transposition of the Great Vessels?

Transposition of the great vessels (TGA) is a relatively rare form of congenital heart disease. It is the second most common cause of a heart defect that can make a baby blue. Transposition means that the great vessels (the Aorta and the pulmonary artery) are “crossed over” or switched so that the aorta arises from the right ventricle and the pulmonary artery arises from the left ventricle. This causes the “blue” or blood without oxygen to be pumped out the Aorta and hence out to the body, and the “red” blood with oxygen to be pumped out to the lungs. This situation, if not corrected, is lethal because the tissue of the body needs to have oxygenated blood to survive. Oxygen is a chemical necessary for the body to “burn” fuel or food to make energy. Sometimes TGA can be associated with other defects of the heart. These include a hole in the wall between the ventricles (VSD), narrowing of the aorta and pulmonic stenosis (a narrowing of the pulmonic valve). These other defects may change how soon these babies are noticed.

Usually TGA is noticed very early after the baby is born because the nurse or doctor will notice that the baby is blue. Frequently it can be diagnosed in utero. Usually, the baby appears quite comfortable and will breathe normally at first and even feed O.K. If the blueness isn't noticed for several hours the baby will usually start to breathe more rapidly. As time goes on the baby will begin to get sick, and breathe more rapidly and not feed well. There is normally an opening between the upper chambers of the heart (atria) when babies are first born. This is called a Patent Foramen Ovale “PFO” the. Also there is a structure call a patent ductus arteriosus (PDA), which is open when a baby is first born. Both of these structures allow for some mixing of blue blood with the red blood so that at least some of the blood going out the aorta has some oxygen in it. Unfortunately, the PDA usually begins to close shortly after the baby is born. Many times the hole between to upper chambers “PFO” is not adequate to provide enough oxygen to the aorta.

Today we have the ability to treat TGA with good results. Surgery is usually done within the first four or five days of life. The babies can usually be stabilized by giving them a medicine called prostaglandin to keep the PDA open. This allows enough oxygen to get to the body. Sometimes it is necessary to do a cardiac catheterization and do what is called a balloon septostomy. This is where a special catheter with a balloon on the end is passed across the hole between the upper chambers of the heart (PFO) and the balloon is pulled across the PFO tearing it to make the hole bigger. This again allows more blood with oxygen to mix with blood without oxygen and allows more red or oxygenated blood to go out the aorta and to the body.

The surgery consists of cutting off the great vessels just after they leave the heart and reversing them so that they are attached to the proper ventricles. This is called the **Jatene** operation. This allows the blood without oxygen to go to the lungs and the blood with oxygen to go to the body. When this operation is done the coronary arteries (blood vessels that feed the heart muscle itself) must be switched so that they are attached to the aorta. Once this operation is done normal circulation is restored, the baby is pink and usually these children can have a pretty normal lifestyle. There can sometimes be problems with kinking of the coronary arteries as the child becomes Older. There are also concerns about the development of early hardening of the coronary arteries.

Another type of surgery that is sometimes done when there are other types of problems with the heart in addition to TGA is called the **Senning** operation. In this case the blood flow from the right atrium is directed to the left ventricle and the flow from the left atrium is directed to the right ventricle. The result of this operation is that blue blood is directed to the lungs and red blood (with oxygen) is directed to the aorta. The disadvantage to this operation is that the right ventricle remains the ventricle pumping blood to the body. This presents a problem because the right ventricle was not “made” to pump at high pressures needed in the aorta and usually these patients will get into trouble in their 20s or 30s.

Great progress has been made in the treatment of TGA. There are still many unknowns and these children will need to be followed closely by a cardiologist for life. They would need SBE Prophylaxis when the need arises.

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