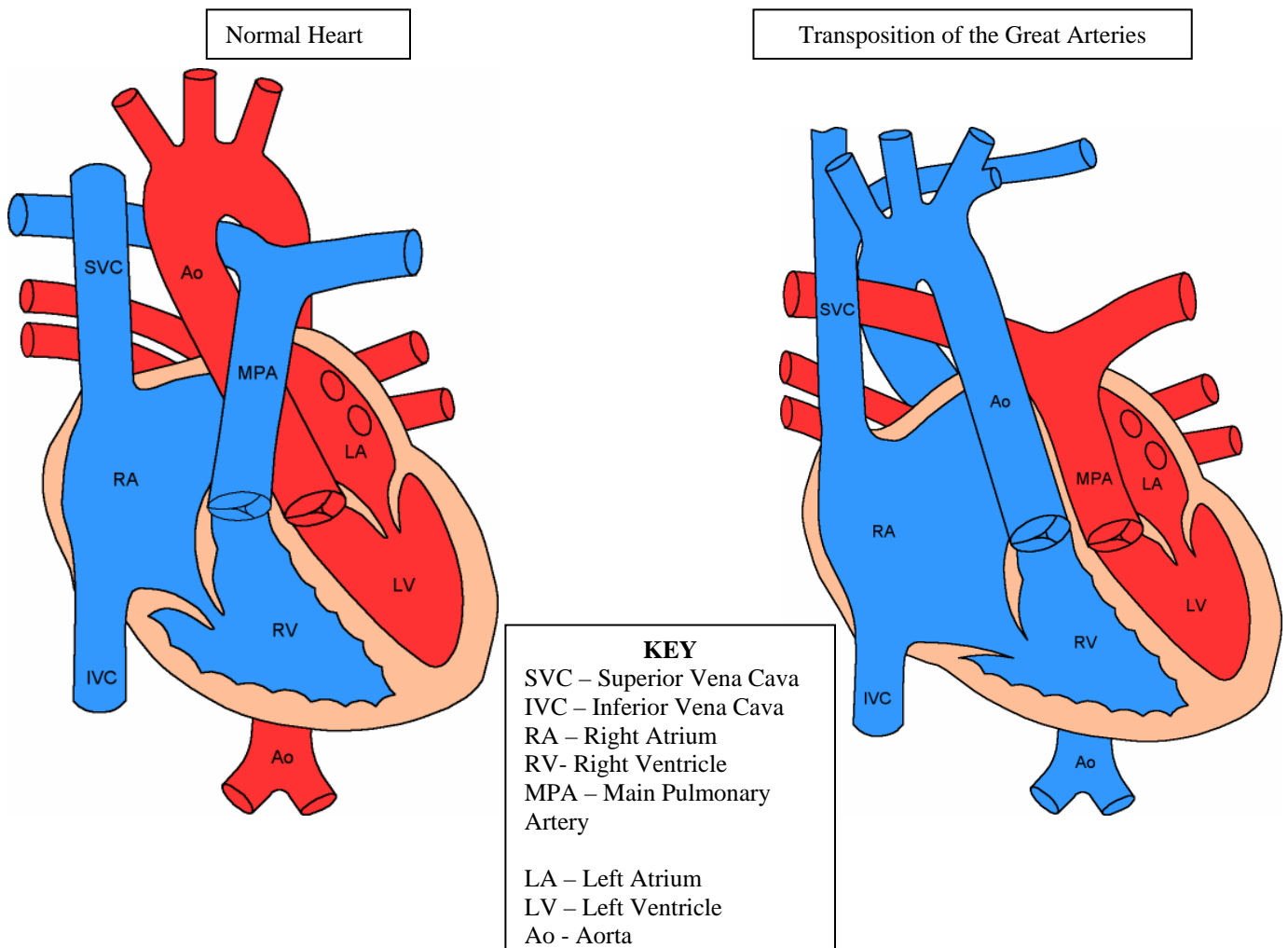


The Adolescent and Adult Congenital Heart Disease Program

The Heart Center at Nationwide Children's Hospital
& The Ohio State University

D- Transposition of the Great Vessels

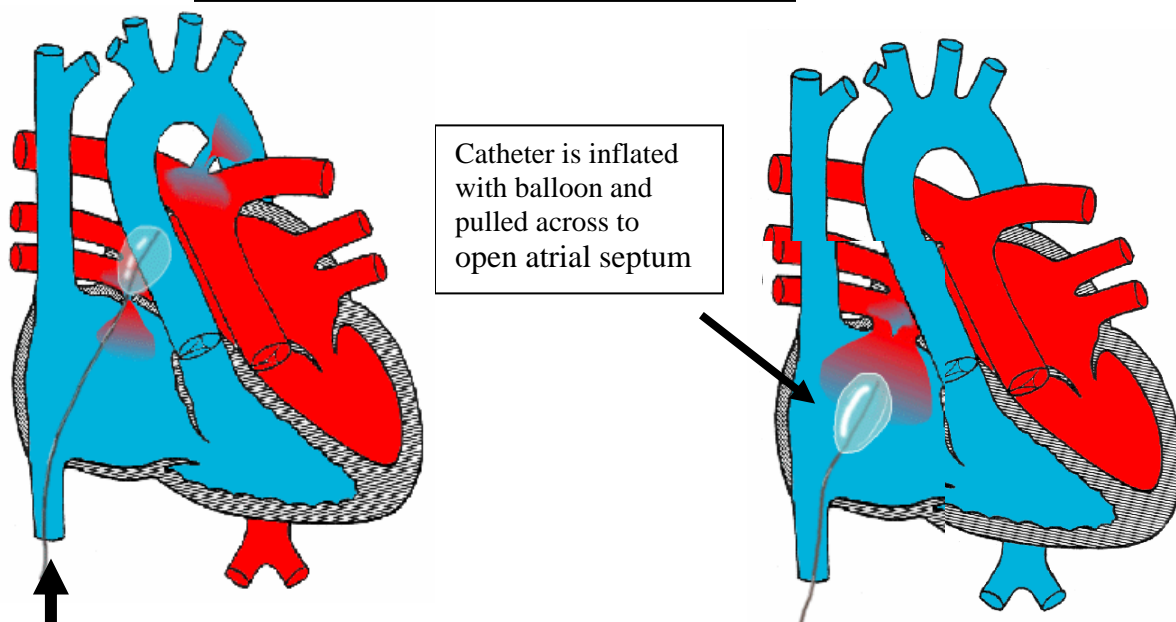
D- transposition of the great vessels (d-TGA) is a congenital heart defect where the aorta and pulmonary artery are switched from their normal positions. In the normal heart, the right ventricle pumps out to the pulmonary artery (to the lungs) and the left ventricle pumps out to the aorta (to the body). Instead, in d-TGA, the aorta comes off of the right ventricle and the pulmonary artery comes off the left ventricle. The ventricles are the pumping chambers for the heart; the left ventricle was created to handle high pressure (out to the body) and the right ventricle was created to handle lower pressure (out to the lungs). In d-TGA, the blue (low-oxygen) blood returning from the body goes right back to the body instead of going to the lungs to get oxygen (to become red blood). The red (with oxygen) blood returns from the lungs and goes right back to the lungs, instead of to the body to give the tissues oxygen.



EARLY TREATMENT (NON-SURGICAL)

Shortly after birth, babies with d-TGA are blue because not enough oxygen is getting to the body. Because of this, many will undergo a cardiac catheterization (heart cath) where a small catheter (tube) is placed in a blood vessel in the groin that tunnels up into the heart. A balloon is contained on the catheter and is inflated and pulled across the wall in the upper chambers (atrial septum) of the heart to create a hole. This way, red (oxygen) blood will mix with the blue (low-oxygen) blood and improve the oxygen level in the body. However, this procedure is only temporary and later requires surgical repair.

Balloon Atrial Septostomy

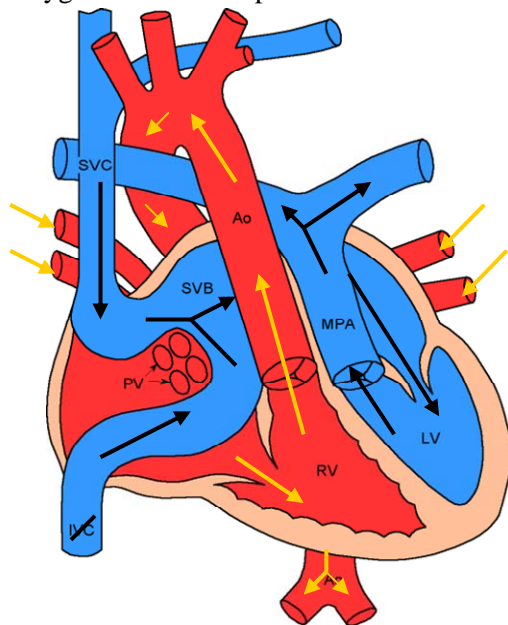


SURGICAL PROCEDURES

Up until approximately 1990, most surviving patients with d-TGA underwent a Mustard operation at around 1-2 years of age. This procedure, named after the surgeon who created it, permanently switches the red and blue blood flow by using “baffles”, typically created out of synthetic material called GorTex. These baffles direct the blood through the atria (upper collecting chambers of the heart) to the proper place. By doing so, blue (low-oxygen) blood is directed to the left ventricle and then to the lungs to pick up oxygen. The red (with oxygen) blood is directed to the right ventricle and then to the aorta and body. After this surgery, the body’s oxygen levels are expected to be normal.

Blue (low-oxygen)
blood returns from
body and travels to the
lungs—(black arrows)

Red (with oxygen)
blood returns from
lungs and travels to the
body and its tissues—
(yellow arrows)



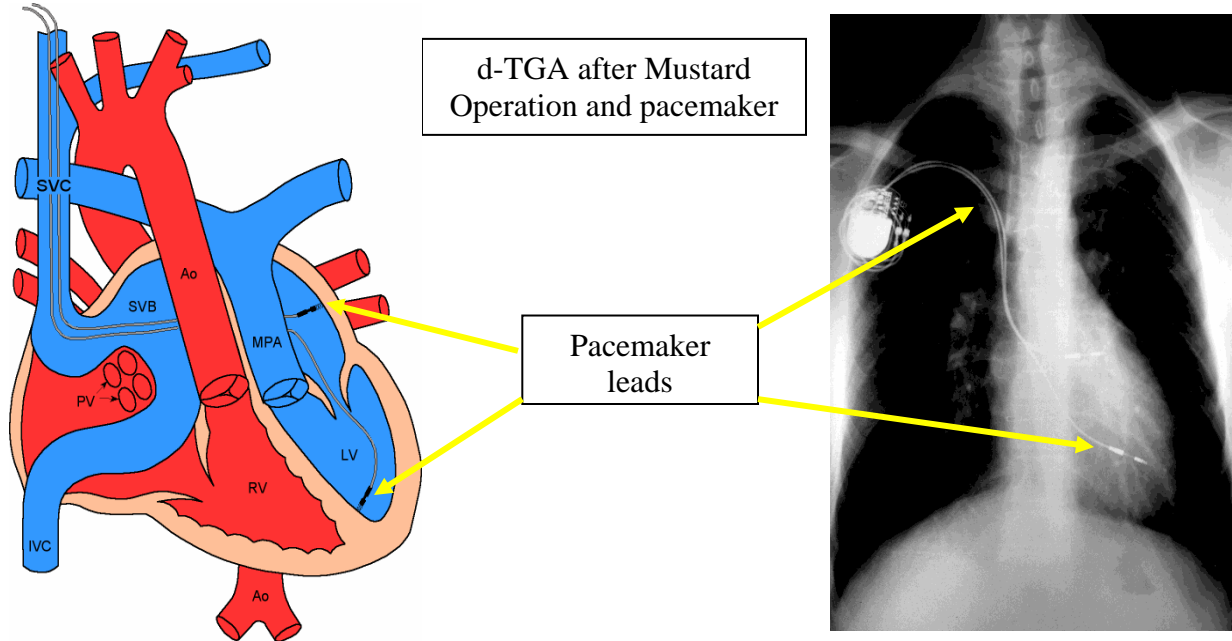
KEY

SVC – Superior Vena Cava
 IVC – Inferior Vena Cava
 SVB- Systemic Venous Baffle
 LV- Left Ventricle
 MPA – Main Pulmonary Artery
 PV- Pulmonary Venous Baffle
 RV- Right Ventricle
 Ao - Aorta

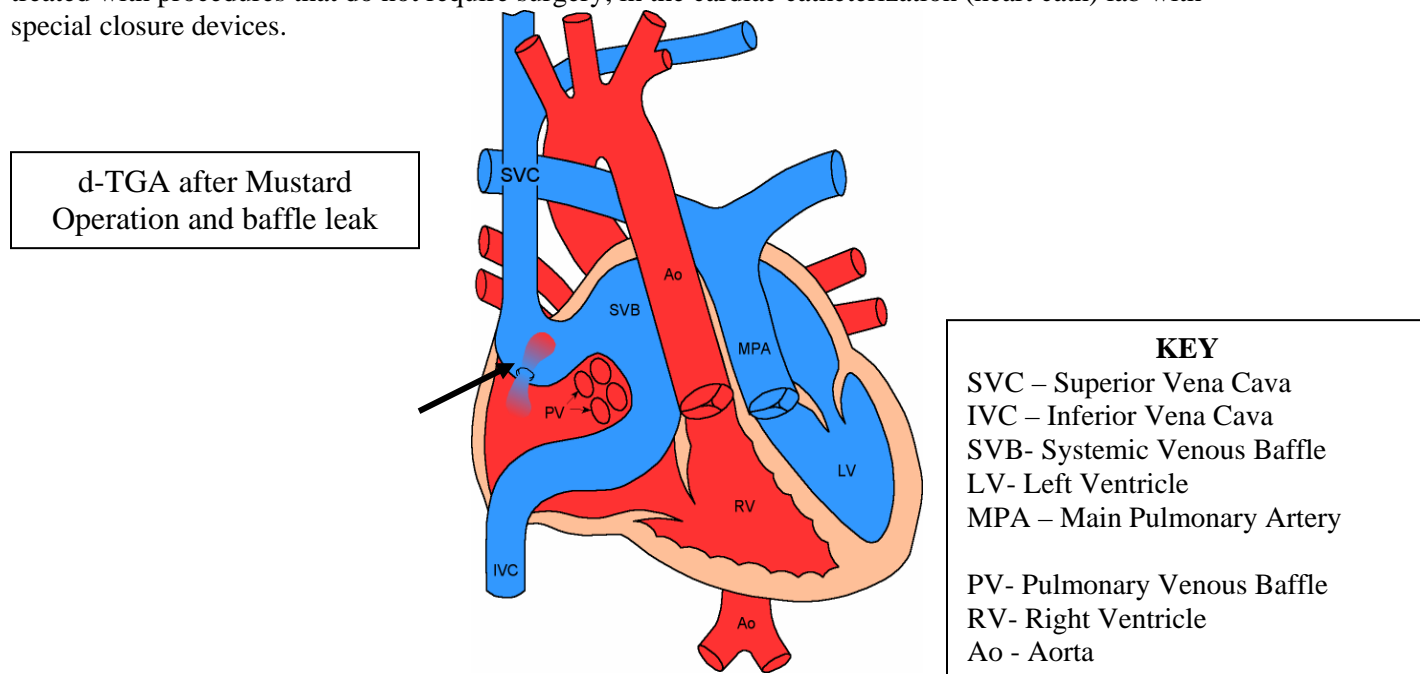
LONG TERM COMPLICATIONS from d-TGA (after Mustard Procedure)

Most patients do extremely well for many years, but as they approach late teenage years and into their 20s, despite previous surgery, some may develop complications such as:

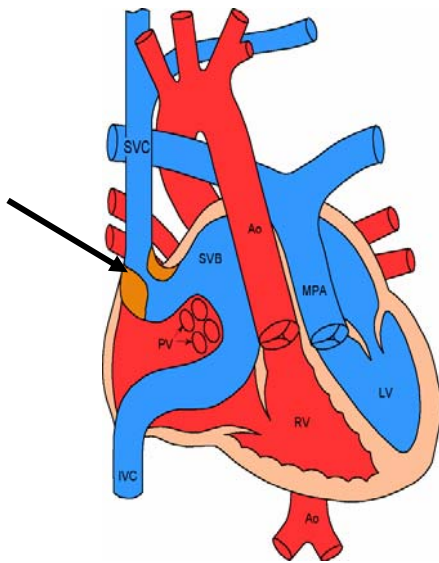
Arrhythmias: The electrical system of the heart runs from the right atrium (upper collecting chamber) and through the wall separating the ventricles (bottom pumping chambers). Due to the extensive stitching in the heart, the heart's electrical system is often damaged, therefore patients may develop slow heart rate, fast heart rhythms, or other abnormal heart rhythms. Depending on the heart rhythm problem, these may require medication or placement of a pacemaker/and or internal cardiac defibrillator (ICD) because some abnormal heart rhythm problems may cause passing out episodes or even sudden death.



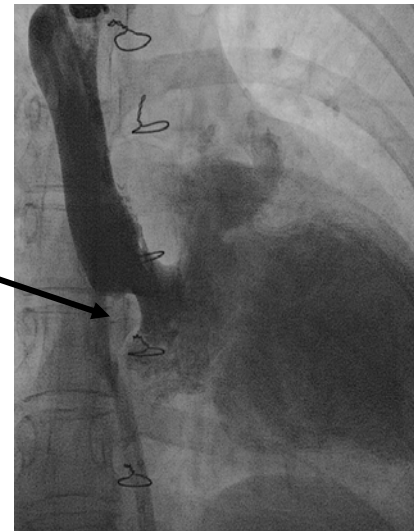
Baffle leaks: Small leaks commonly occur over time in the Mustard baffles and cause lower oxygen in the body (blueness or cyanosis) when blue blood again mixes with the red blood and goes out to the body. They may occur in the baffles that supply blood to the lungs (systemic venous baffle) or that supplying blood to the body (pulmonary venous baffle). More concerning, is the risk for stroke or “mini-strokes” if blood clots cross this leak and enter the arteries going to the brain. For most patients, baffle leaks can be treated with procedures that do not require surgery, in the cardiac catheterization (heart cath) lab with special closure devices.



Baffle Obstruction: The baffles, that were created to switch the blood flow in the heart, are usually made from a synthetic material, called GorTex. With time, the baffles can scar which causes difficulty with blood flow (obstruction) in the heart. The most common site of obstruction is where the head and neck vein (superior vena cava) comes into the heart. Also, obstruction can occur at the point where the pulmonary veins (from the lungs) enter the heart. Baffle obstruction can cause symptoms such as shortness of breath. In most situations, baffle obstructions are diagnosed by echocardiography (heart ultrasound), however, heart MRI or catheterization may be necessary diagnostic tests to detect baffle obstruction. If the obstruction is in the systemic venous (SV) baffle, it can often be alleviated in the cardiac catheterization lab using transcatheter stent therapy with a combination of balloon angioplasty and transcatheter stent therapy. A stent is a mesh-like metal tube that is loaded into a catheter (or small tube) and is inserted into an artery in the groin. The catheter is advanced into the heart, at the area of obstruction, and a balloon is inflated causing the stent to expand and open the area of narrowing. In cases of pulmonary venous (PV) baffle obstruction, surgery is typically necessary to alleviate the narrowing.

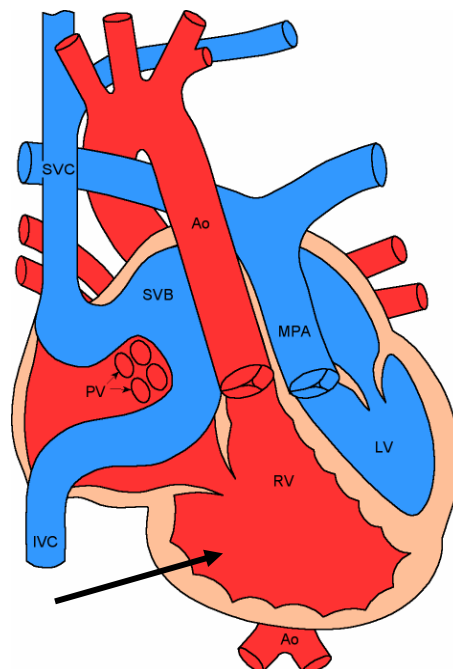


d-TGA after Mustard Operation and SV baffle obstruction



Heart Failure: Although the heart has been repaired, it is still not normal. The right ventricle, which usually pumps blood to the lungs (low-pressure) now pumps blood to the body (high pressure). The right ventricle handles the high-pressure load fairly well for many years but, eventually begins to wear down. Eventually, it becomes dilated, boggy, and weakens. This weakening is can cause symptoms of heart failure such as shortness of breath, tiredness, chest pain, and decreased activity ability. Treatments with medicines are typically helpful and can restore some of the heart function and improve symptoms. If the function of the right ventricle continues to worsen, despite medical treatment, heart transplant is considered.

d-TGA after Mustard Operation and right ventricular enlargement



KEY

- SVC – Superior Vena Cava
- IVC – Inferior Vena Cava
- SVB- Systemic Venous Baffle
- LV- Left Ventricle
- MPA – Main Pulmonary Artery
- PV- Pulmonary Venous Baffle
- RV- Right Ventricle
- Ao - Aorta

KEEP YOURSELF HEALTHY:

- Maintain routine cardiac follow-up
- Notify your cardiologist of concerning symptoms
- Take medications as they are prescribed (including antibiotics for dental procedures, tattoos, & piercings if indicated for your heart defect)
- Maintain a healthy weight
- Eat a healthy diet (low-sodium, low-fat)
- Avoid all tobacco products
- Participate in regular aerobic exercise (walking, swimming, jogging, or biking) for at least 30 minutes, most days of the week. Patients should avoid isometric activities, or those to the point of grunting or straining (chin-ups, push-ups, heavy lifting) as these activities cause added strain to the heart
- Avoid dietary or medication sources of stimulants (caffeine in soda or chocolate, ephedrine/pseudoephedrine-containing decongestant cold medications)
- If you are female, it is important to plan pregnancy with your cardiologist to ensure optimal cardiac status, prior to becoming pregnant