Review Article

Transposition of the Great Arteries: Diagnosis, and Treatment

Abstract

Background: Transposition of the great arteries (TGA) is the most common cyanotic congenital heart disease that manifests itself in newborns. A distinctive feature of Transposition of the great arteries is ventriculoarterial dissonance, in which the aorta arises from the morphologically right ventricle and the pulmonary artery from the morphological left ventricle. In approximately one-third of patients with major arterial transplantation, the anatomy of the coronary artery is abnormal: left coronary artery, circling the left coronary artery, exiting the right coronary artery (22%), single right coronary artery (9.5). (%), Left coronary artery only. The arteries (3%) or the reverse origin of the coronary arteries (3%), represent the most common types.

Conclusion: If there are no unusual risk factors, more than 98 percent of surgically treated infants live in infancy. Most children who undergo TGA surgery recover and grow normally, although they may have little risk in the future for arrhythmias, leaky valves, and other heart problems.

Keywords: *Ventricular Septal Defect; Left Ventricular Outflow Tract; Great Artery; Arterial Switch Operation; Pulmonary Artery.*

Introduction

Transposition of the great arteries is a serious but rare heart defect that occurs at birth (congenital), in which the two main arteries from the heart are reversed. This condition is also known as dextro-transposition of the great arteries. A rare variant of this condition is called Levo-transposition of large arteries. Enlargement of the large arteries alters blood circulation in the body, leading to a lack of oxygen in the blood that flows from the heart to the rest of the body. Without an adequate supply of oxygen-rich blood, the body cannot function properly and the child faces serious complications or death without treatment. Displacement of the large arteries is usually detected in the weeks from prenatal or first hours of life. Corrective surgery after birth is the most common treatment. Having a baby with transposition of the great arteries is risky, but with the right treatment, perspective is promising (1).

Causes and Risk Factors

While TGA is developing, It is unknown at this time what he will do after leaving the post. Typically, the pulmonary artery, which carries blood from your heart to your lungs to receive oxygen, is connected to the lower right chamber (right ventricle). From the lungs, oxygenated blood travels to the upper left chamber (left atrium) of your heart, through the mitral valve to

the lower left chamber (left ventricle). The aorta is usually connected to the left ventricle. This oxygen-rich blood carries from your heart to the rest of your body. In the transposition of great arteries, the positions of the pulmonary artery and the aorta change. The pulmonary artery is connected to the left ventricle and the aorta is connected to the right ventricle (2).

Low oxygen blood circulates in the right side of the heart and returns to the body without passing through the lungs. Oxygen-rich blood circulates in the left side of the heart and returns to the lungs without circulating to other parts of the body. When low-oxygen blood circulates in the body, it produces cyanosis on the skin. For this reason, transposition of large arteries is called cyanotic congenital heart disease. Various factors such as genetics, rubella, other viral diseases in pregnancy, diabetes in mothers over 40 years of age, or their mothers may increase the risk of this condition, but in most cases the cause is unknown (2).

Levo-Transposition: In this rare type of transposition, the left artery of the large artery, or congenitally corrected transfusion, two ventricles are placed so that the left ventricle is on the right-side heart. Can be changed. It receives blood from the right ventricle on the left side of the heart and blood from the left ventricle. This type of transposition of the Great Arteries is also called Congenital Corrective transposition. However, blood usually circulates properly in the heart and body. The left ventricle attaches to the pulmonary artery and sends oxygen-poor blood to the lungs, and the right ventricle on the left attaches to the aorta and carries oxygen-rich blood to the body. Due to congenital correction of circulation, some people with this rare condition will not experience symptoms for years and will not be diagnosed until puberty. However, many people with this condition have other congenital heart diseases that cause symptoms and these people often suffer from heart disease in childhood. Depending on your condition and the defect in your heart, you may have surgery to correct the defect (3).

The right ventricle can eventually cease to function as the main pump chamber, which can lead to heart failure in adults. In adults, the valve that protects the right ventricle (tricuspid valve) often does not function properly. Valve and ventricular dysfunction can lead to heart failure. Ultimately, treatments such as valve replacement, ventricular assist devices, and heart transplants may be required. Risk factors: The exact cause of the aortic obstruction is unknown, but several factors may increase the risk of giving birth to a baby in this condition, such as rubella or another maternal viral disease with a history of pregnancy, Drinking during pregnancy, smoking during pregnancy, mothers, with poor control of diabetes (3).

Pathophysiology of TGA

The systemic and pulmonary circulation is completely different. Upon returning to the right heart, the systemic venous blood is pumped to the lungs in a systemic circulation without oxygen. The oxygenated blood that goes to the left heart returns to the lungs instead of going to the rest of the body. This difference is incompatible with life unless unsaturated and oxygenated blood merges in one or more pathways (e.g., an arterial, ventricular, or large arterial pathway) (4).

Types of TGA

Atherosclerosis may occur in the transposition of vessels. Effects can range from minor changes in blood pressure to circulatory disorders, depending on the type and height of the graft and the individual vessels involved. Although "transposed" literally means "changed" many types of TGVs have unusually positioned vessels that are not interconnected. The terms TGV and TGA are often used for dextro-TGA, where the two main arteries are in an altered position. However, for Levo-TGA in both arteries and ventricles, both terms are often used somewhat interchangeably; Other errors in this category cannot be identified by any of these terms (5).

Dextro-transposition of the great arteries of the aorta is a cyanotic heart disease in which the aorta originates in the right ventricle and the pulmonary artery originates in the left ventricle. This switch immediately pumps unoxygenated blood from the right heart into the aorta, completely bypassing the lungs and circulating around the body and heart. In this condition, the left heart does not return to the body as usual, but continuously returns oxygen-rich blood to the lungs through the pulmonary arteries. The result is two separate "parallel" systems. This is called cyanotic congenital heart disease (CAD) because the newborn becomes blue (cyanosis) due to a lack of oxygen (figure 1) (5).

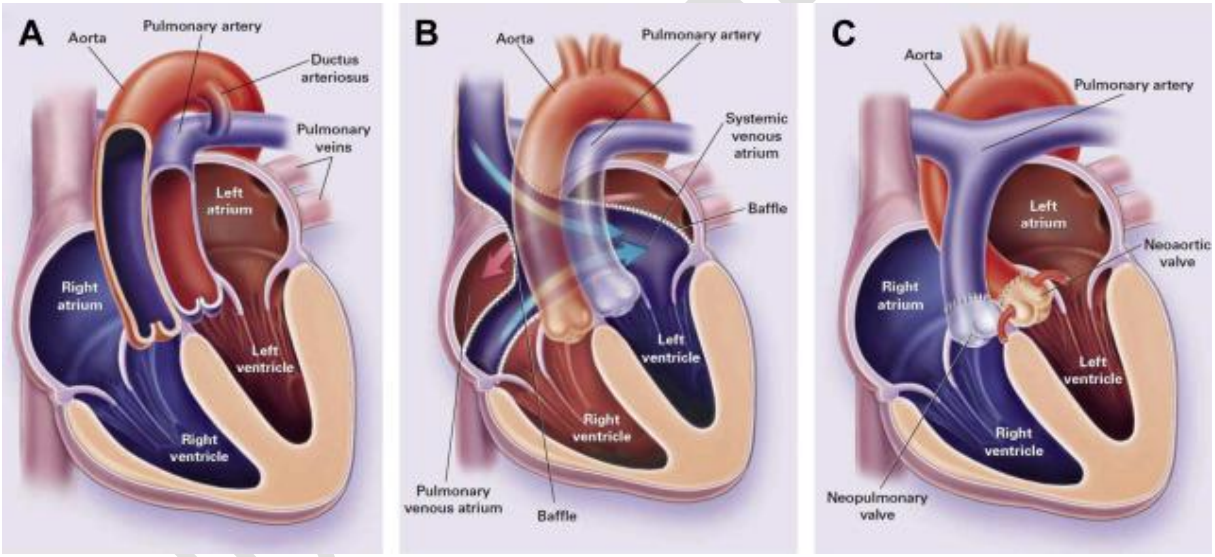

Figure 1 Dextro-Transposition of the Great Arteries (5)

Levo-Transposition of the great arteries is a rare heart disease in which the main arteries move to the left of the anterior aorta. The pulmonary artery and the left and right ventricles also move with their corresponding atrioventricular valves. In other words, the right ventricle is on the left side of the heart and the left ventricle is on the right side of the heart. This condition involves low, systemic blood flow. Despite its high resistance to the systemic circulation, the right ventricle, which favors the pumping of blood to the pulmonary circulation at low pressure, can lead to complications due to pressure changes, since it has to pump blood at very high pressures. Because now it is in the position in which the left ventricle normally is (figure 2)(6).

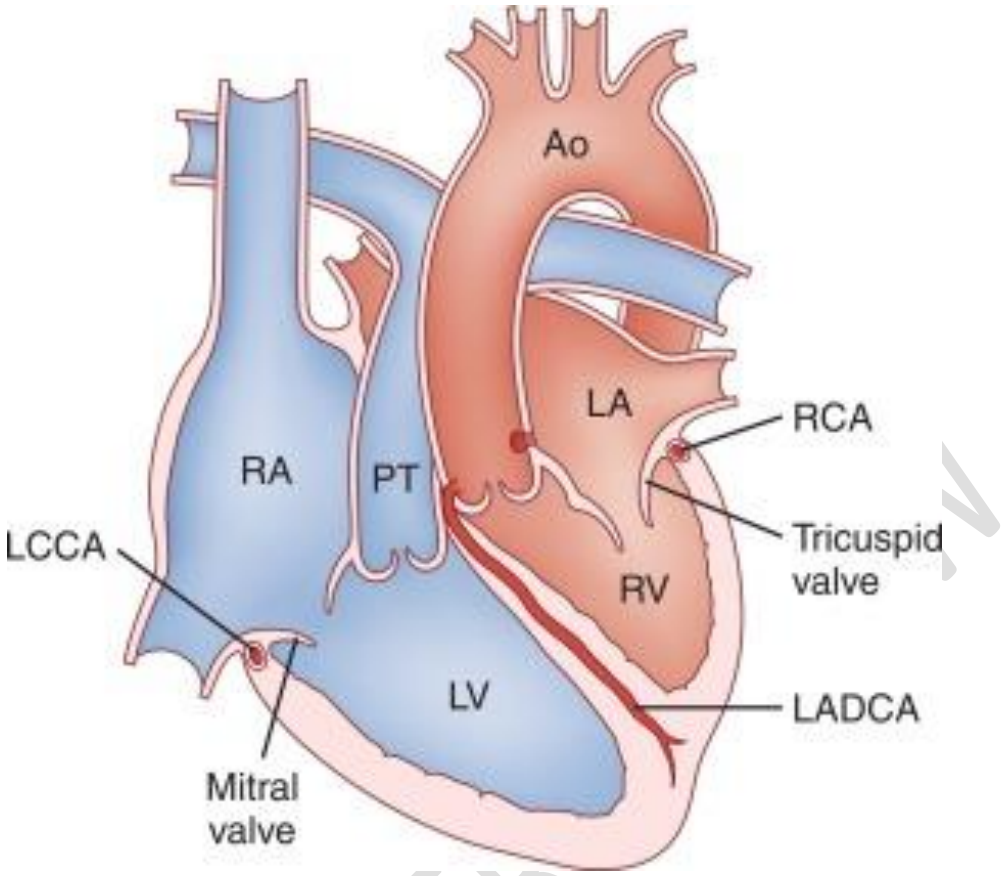

Figure 2 Levo-Transposition of the great arteries (6)

Simple and complex TGV: TGV is commonly associated with other heart defects, and the most common types of intracardiac shunts are the patent foramen ovale, ventricular septal defect, patent ductus arteriosus.If patients have atrial septal defect, There may also be narrowing or other defects in the valves and/or blood vessels. If there are no other heart defects, it is called "simple" TGV. If there are other defects, it is called "complex" TGV (7).

Complications

Possible complications of a large artery transposition include oxygen deficiency in tissues: baby tissues get too little oxygen (hypoxia). If your child does not have a mixture of oxygenated blood and unoxygenated blood, he will not survive. A heart attack is a condition in which the heart cannot pump enough blood to meet the body's needs, and the right ventricle pumps at a higher pressure than usual and develops over time. This extra pressure can harden or weaken the right ventricular muscles. Damage to the Lung will cause shortness of breath. Early in life, usually in the first week, all infants with large arterial implants need surgery. The most common type of surgical arterial switch is the operation to correct the transposition when identified in children. During this operation, the surgeon moves the large arteries so that they are properly connected to the pumping chamber. The heart supply arteries (coronary arteries) must also be replaced. Although this operation is life-saving, it can cause problems in life, including narrowing of the arteries that supply blood to the heart (coronary arteries), heart rhythm abnormalities (arrhythmia), myocardial infarction can lead to heart failure, and narrow vessels connecting large vessels and leaky heart valves (8).

Prevention

In most cases, the transmission of large arteries cannot be prevented. If patients have a family history of heart defects or have a child with congenital heart failure, patients should speak to a genetic counselor and cardiologist who had congenital heart defects before pregnancy. In addition, it is important to take steps to have a healthy pregnancy. For example, before you become pregnant, start taking a multivitamin that contains 400 micrograms of folic acid (8).

Symptoms

Severe cyanosis occurs within hours of birth, followed by metabolic acidosis, which is due to poor tissue oxygenation. Patients with moderate or severe atrial septal defect, a ventricular septal defect, a patent ductus arteriosus, or a combination may have symptoms and signs of heart failure (e.g., tachypnea, dyspnoea, tachycardia, sweating, Inability to gain weight) can develop in the first few weeks of life. Other than generalized cyanosis, the physical exam is not significant. There cannot be a heart murmur unless there are abnormalities associated with it. The second heart sound (S2) is loud (9).

Diagnosis

TGA in infants can be diagnosed before birth, but it can be difficult to diagnose. The prenatal test is usually not done to see how large the arteries become until the doctor suspects that the baby may have congenital heart disease. After the baby is born, if the baby's skin turns blue or has difficulty breathing, doctors immediately suspect a heart problem, such as a TGA. If a child has another heart defect, such as the blue color of the skin that may not be noticeable. If the hole is in the upper cavity of the heart, it is called atrial septal defect. This disorder in the lower chambers of the heart is called ventricular septal defect. It is also possible that the child has patent ductus arteriosus. It is the opening between the two large blood vessels of the heart, the aorta, and the pulmonary artery, where oxygen-rich blood and unoxygenated blood can meet (10).

As your baby becomes more active, heart defects do not allow enough blood to flow, cyanosis becomes apparent. The child's doctor may also suspect heart failure if he hears a heartbeat, which is an abnormal wheezing sound caused by impaired blood flow. Physical examination is not sufficient to accurately detect large artery transposition. Accurate diagnosis requires one or more of the following tests (10).

Echocardiogram

An echocardiogram is an ultrasound scan of the heart: it uses sound waves arising from your baby's heart and creates moving images that can be viewed on a video screen. Doctors use this test to detect large artery translocations based on the condition of the aortic and pulmonary arteries. An echocardiogram may also reveal cardiac defects such as ventricular dysfunction, atrial septal defect, or ductus arteriosus (figure 3) (11).

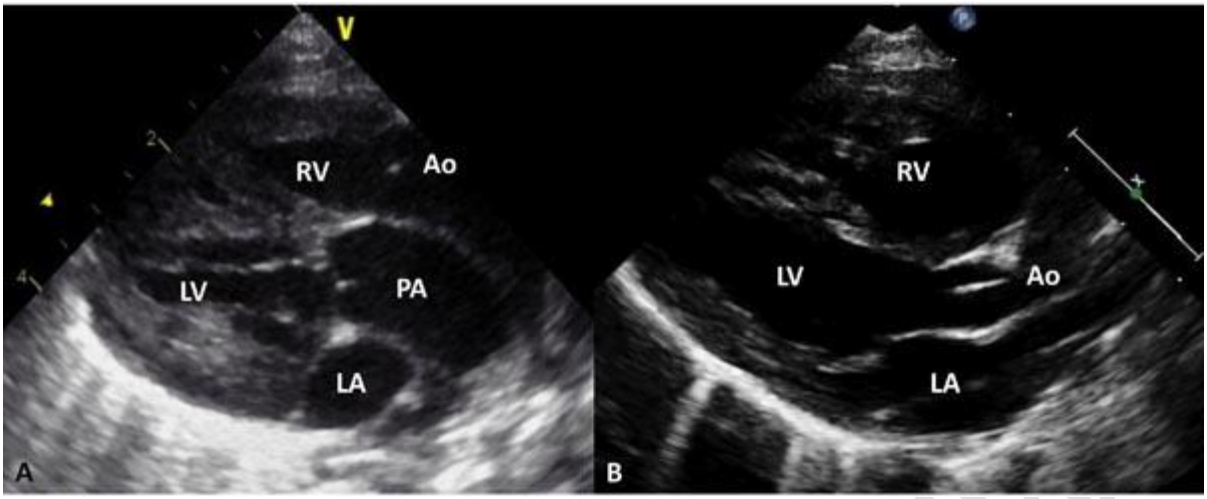

Figure 3 Echocardiogram of Transposition of the Great Arteries (A), Normal (B) (11)

Cardiac catheterization

This procedure is usually done only when other tests, such as an echocardiogram do not provide sufficient information for diagnosis. During cardiac catheterization, the doctor inserts a thin, flexible tube (catheter) into the artery or vein of the baby's groin and carries it to his or her heart. The dye is injected through a catheter so that the baby's heart structures can be seen on an X-ray. The catheter measures the pressure in heart chambers and blood vessels and can measure the amount of oxygen in the blood. Cardiac catheterization can be performed to treat the temporal displacement of the immediate large arteries (balloon atrial septostomy) (figure 4) (12).

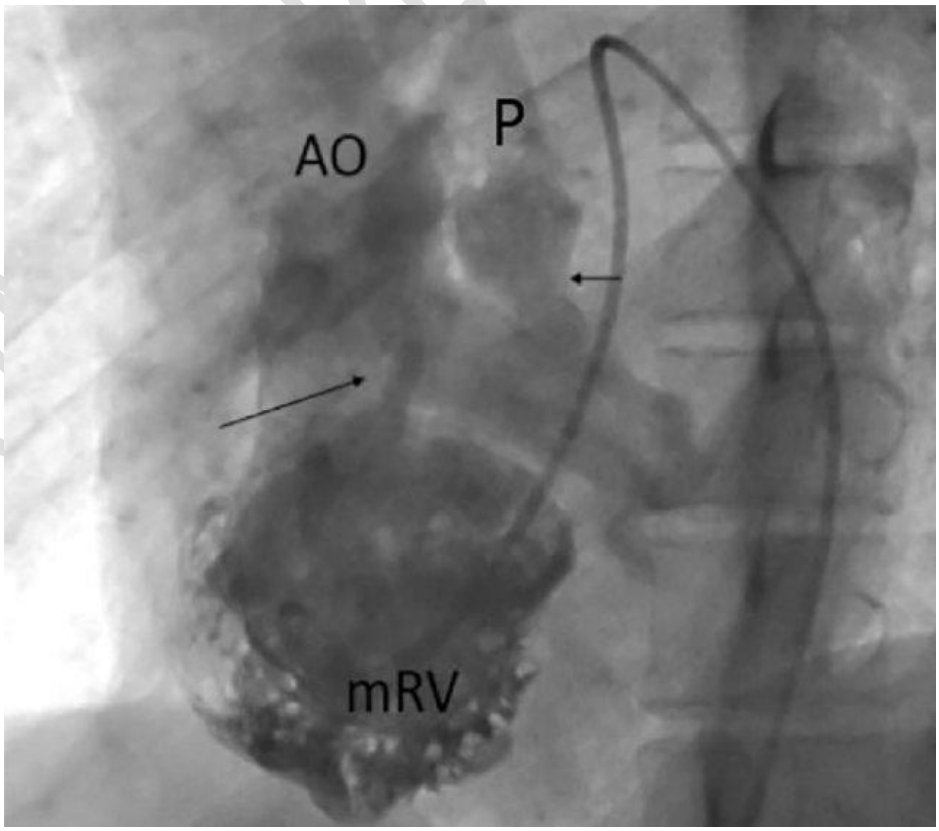

Figure 4 cardiac catheterization of a CCTGA and Situs Inversus patient showed the anteriorly placed aorta arising from the morphologic right ventricle and the posteriorly placed pulmonary artery arising from the morphologic left ventricle and in association with ventricular septal defect (long arrow), pulmonary stenosis (short arrow) (12)

Chest X-ray

Although a chest x-ray does not provide an accurate diagnosis of a large artery transposition, it does allow the doctor to see the size of the baby's heart and the condition of the aorta and pulmonary artery (figure 5) (13).

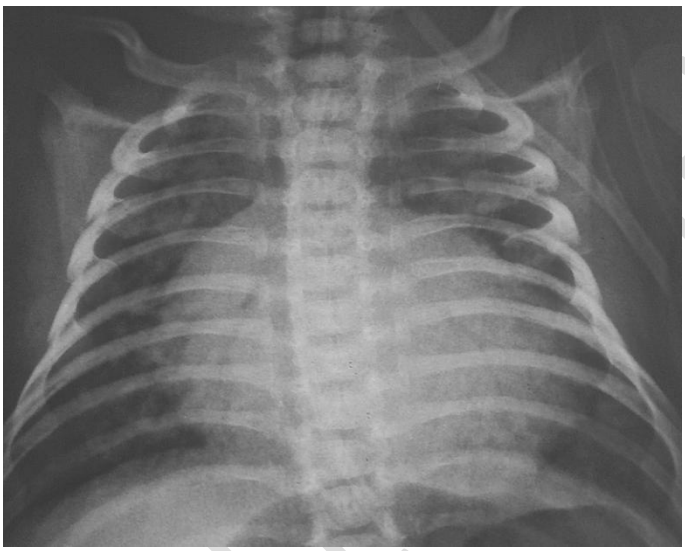

Figure 5 Chest X-ray of Transposition of the Great Arteries (13)

Electrocardiogram

An electrocardiogram records the electrical activity of the heart with each beat. During this procedure, patches with wires (electrodes) are applied to the baby's chest, wrists, and ankles. Electrodes measure electrical activity which is recorded on paper (Figure 6) (14).

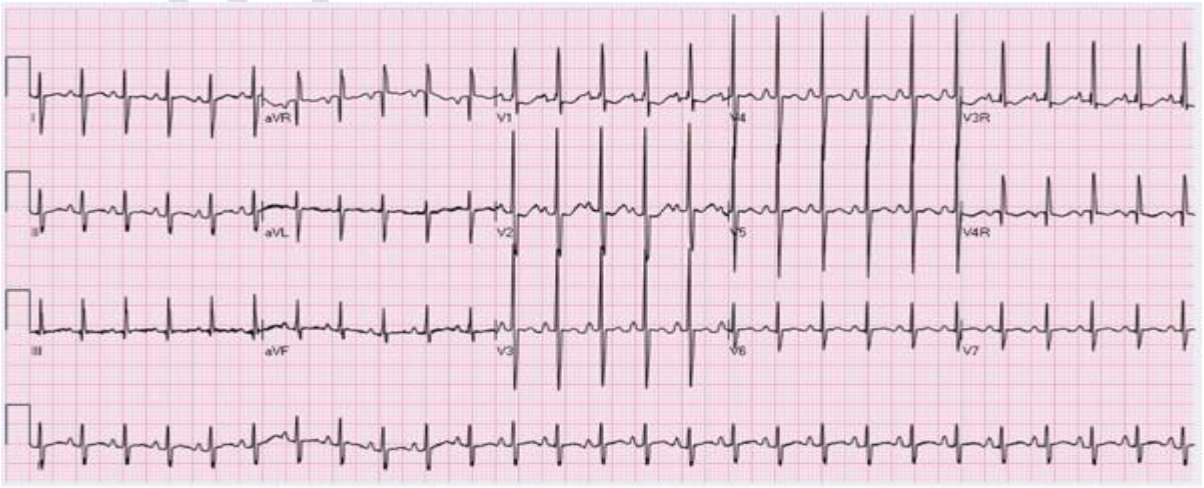

Figure 6 Electrocardiogram of Transposition of the Great Arteries (15)

Treatment

Before surgery

A cardiologist can suggest several options to help manage the condition before corrective surgery. Drug: Prostaglandin E1 helps keep the connection between the aorta and pulmonary artery (ductus arteriosus) open, improves blood circulation, and improves oxygen-rich blood mixing. Atrial septostomy: This procedure is usually performed by cardiac catheterization rather than surgery, which improves the natural connection between the upper chambers of the heart. This allows oxygen-rich blood and oxygen-deprived blood to mix and allows a better supply of oxygen to the baby's body (16).

Surgery

The surgical procedure depends on the age of the patient at presentation, the presence of associated congenital cardiac lesions, and the experience of the cardiothoracic surgeon with a specific surgical technique. Most term newborns with uncomplicated TGA can undergo an artery switch procedure in an operation with minimal mortality (figure 7) (17).

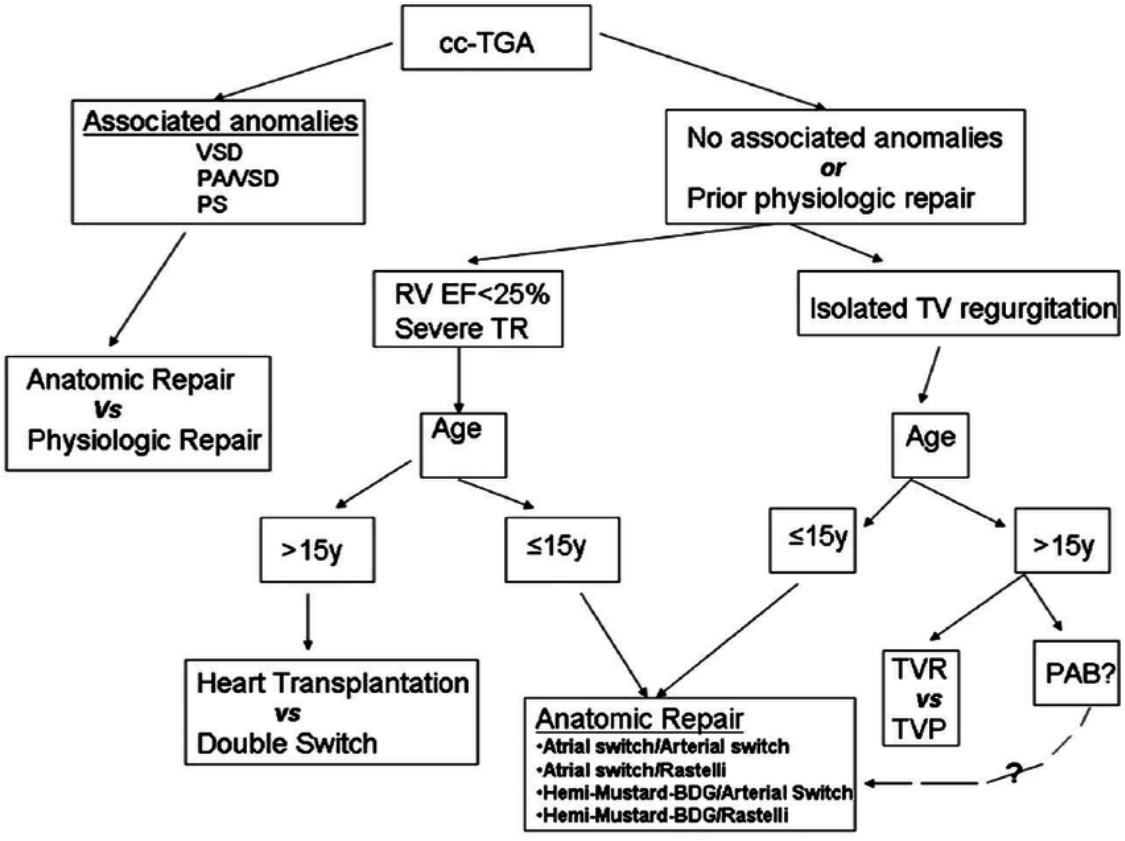

Figure 7 Decision making in choosing the type of surgery, cc-TGA: congenitally corrected transposition of great arteries, BDG: bidirectional Glenn, EF: ejection fraction, PA: pulmonary artery, PAB: pulmonary artery banding, PS: pulmonary stenosis. RV: right ventricle, TR: tricuspid regurgitation, TV: tricuspid valve, TVP: tricuspid valvuloplasty, TVR: tricuspid valve replacement, VSD: ventricular septal defect (18)

Transposition of the great arteries with intact ventricular septum

The ideal surgery is artery switch surgery. It represents anatomical improvement and establishes ventriculoatrial compliance. This procedure should be performed when the baby is less than 4 weeks of age because the left ventricle may not be able to handle the systemic pressure in the low-pressure, low-resistance pulmonary circuit for very long periods. However, in rare cases, depending on the specific anatomy of the coronary artery (eg, intramural coronary artery), dissection of the coronary artery may not be possible and arterial switching is not recommended. In this subgroup, changes in atrial levels appear to reduce short-term and post-surgical morbidity and mortality (figure 8) (19).

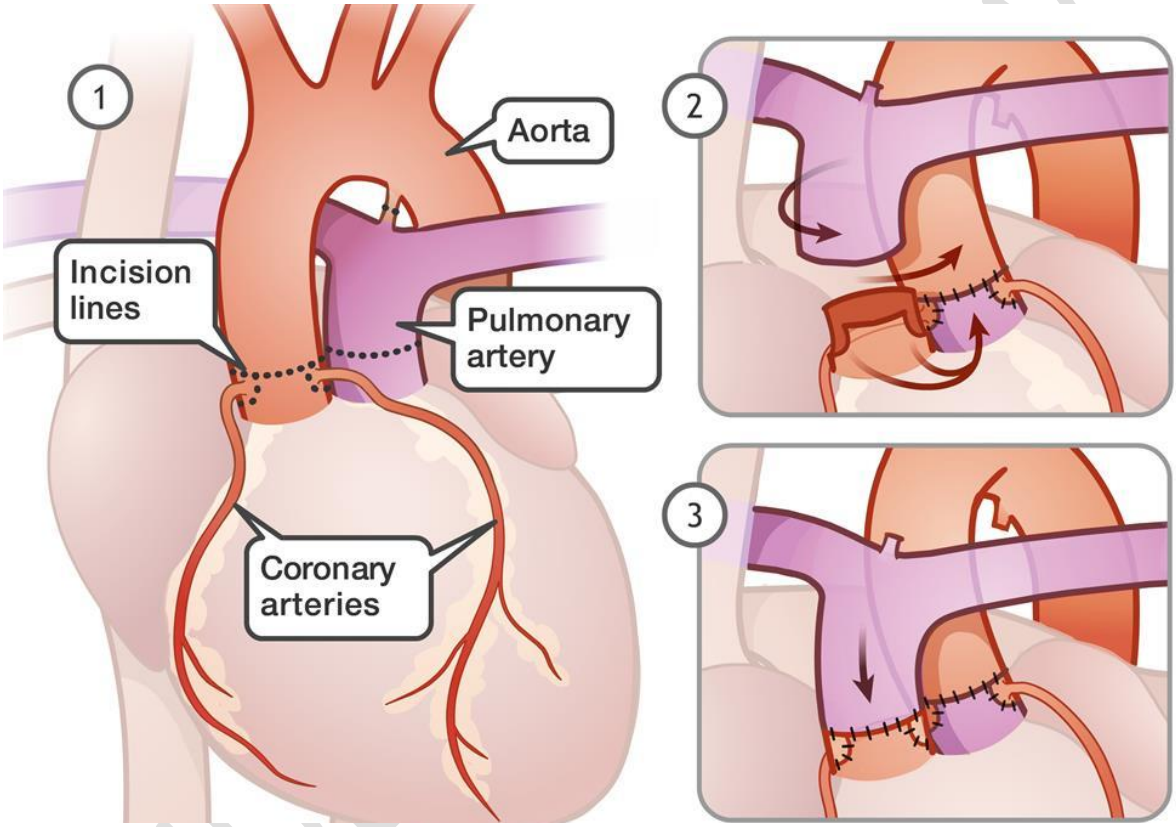

Figure 8 Transposition of the great arteries with intact ventricular septum (19)

Transposition of the great arteries with ventricular septal defect

The preferred procedure is the arterial switch procedure with interventricular septal closure. If the ventricular septal defect is large and complex, and the anatomy of the coronary artery renders the artery switch operation unusable, Rastelli-type intracardiac repair is possible. With a Rastelli-type procedure, it may be better to wait until the child is older, as the right ventricular pulmonary artery needs to be removed during Rastelli surgery. If a child has severe congestive heart failure (with growth retardation), the child may be advised to continue with reconstructive surgery or, if this is not possible, procedures to restrict pulmonary blood flow (figure 9)(20).

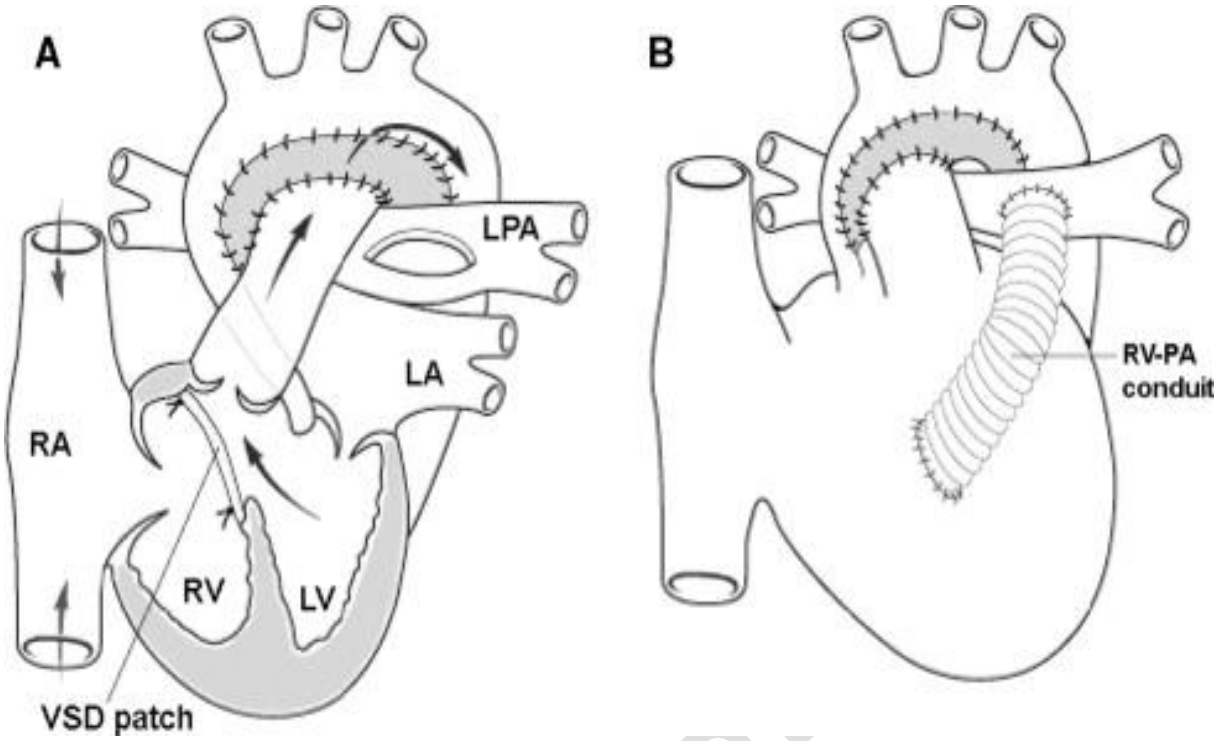

Figure 9 Rastelli procedure (20)

Transposition of the great arteries with ventricular septal defect and left ventricular outflow tract obstruction

Stenosis or occlusion of the pulmonary valve (left ventricular outflow tract) can prevent an arterial switch operation. Rastelli intracardiac repair is possible if the ventricular septal defect is not restricted and is removed from the aorta. Because the Rastelli procedure requires a channel from the right ventricle to the pulmonary artery, it may be wise to postpone recovery until the child is grown. In this case, it may be necessary to introduce an aortic-pulmonary artery bypass during the neonatal period to establish adequate pulmonary blood flow during the waiting period (21).

Transposition of the great arteries with ventricular septal defect and pulmonary vascular obstructive disease

These patients may not be good candidates for surgery due to the progressive increase in pulmonary vascular resistance. This is a small subgroup of patients whose condition is often not diagnosed until palliative or curative treatment is administered (21).

Atrial switch operation

During this operation, the surgeon creates a tunnel (septum) between the two upper chambers of the heart (atria). This moves deoxygenated blood to the left ventricle and pulmonary artery, and oxygen-rich blood to the right ventricle and aorta. In this procedure, as in a normal heart, the right ventricle has to pump blood not only to the lungs but also to the body. Possible complications of atrial switch surgery are arrhythmias, septum obstruction or leakage, and heart failure due to long-term problems with right ventricular function (figure 10)(21).

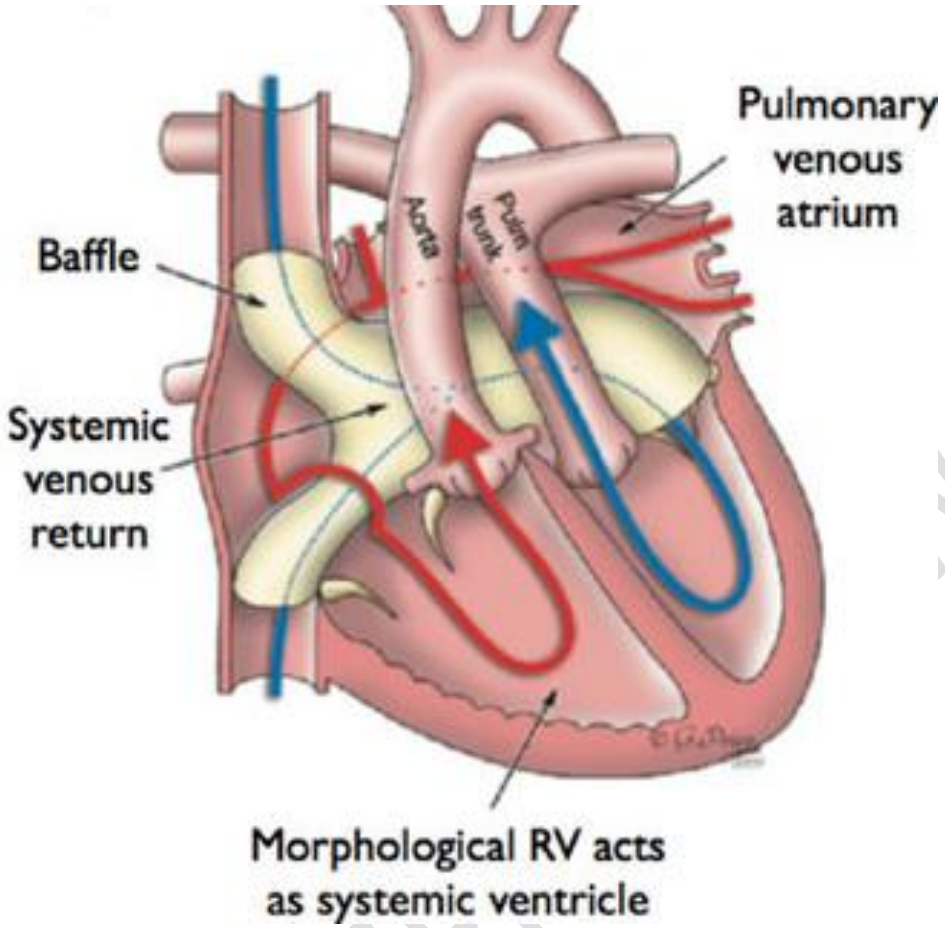

Figure 10 atrial switch operation (Mustard/Senning) for transposition of the great arteries. Systemic (blue) blood is directed from the superior caval vein and inferior caval vein into the left atrium, then via the mitral valve to the left ventricle and then to the pulmonary artery. Pulmonary (red blood) is directed from the pulmonary veins to the right atrium, then via the tricuspid valve to the aorta (21)

After surgery

After corrective surgery, the baby will need lifelong follow-up care from a cardiologist who specializes in congenital heart disease to monitor their heart health. Cardiologists may recommend that the child avoid certain activities, such as weightlifting or competitive sports, as these can raise blood pressure and put pressure on the heart. Many people who undergo artery switch surgery may not need additional surgery. However, some complications such as arrhythmias, heart valve leakage, or heart-pumping problems may require additional treatment (22).

Long-term care

The baby needs lifelong monitoring and treatment. The child will be monitored and regular follow-up visits with a pediatrician will be made. As the baby gets older, he or she will be cared for by a congenital heart specialist who can monitor their condition over time. A large artery transplant can affect a child's adult life, as it can lead to other health problems. Adults with congenital heart defects may need other treatment for their condition (22).

Pregnancy

If transposition of great arteries is repaired in infancy, a healthy pregnancy is possible but may require specialized treatment. If the patient is planning to become pregnant, consult a cardiologist or obstetrician before conceiving. If the patient develops complications such as arrhythmias and severe myocardial problems, pregnancy can put her and her baby at risk. In some situations, for example, in women with serious complications due to a heart defect, pregnancy is not recommended even with transposition repair (22).

Discussion

Many patients with ventricular arterial incompatibility survived to adolescence. People with complete transposition of large arteries often undergo arterial switching (mustard or sensing surgery), in which morphological right ventricle (RV) supports systemic circulation. RV regurgitation and tricuspid regurgitation are common. Some patients may eventually need a heart transplant. Sinus node dysfunction is becoming more common with prolonged follow-up and some patients need a pacemaker. Arterial rhythm is normal. Patients with arterial switching also survive adolescence. Long-term problems include coronary artery stenosis, pulmonary artery dysfunction, and aortic regurgitation. Patients with congenital transplant correction have atrioventricular and ventricular arterial degeneration and thus have morphological RV and thin tricuspid valves in the systemic circulation. Most patients have associated defects such as tricuspid valve abnormalities, ventricular septal defects, and pulmonary stenosis. Heart block occurs with age (23).

Atrial arrhythmias occur frequently, and their presence should facilitate the investigation of hemodynamic problems. Progressive tricuspid regurgitation occurs with age and is associated with impaired right ventricular function. Surgical treatment should be considered at the first signs of right ventricular dilation or dysfunction. All patients who specialize in clinical evaluation, imaging, and hemodynamic evaluation of congenital heart disease in adults should visit the center from time to time. Many patients with the transposition complex survived to adulthood. People who have undergone a complete transposition have been exposed to previous repairs and are increasingly being seen in cardiovascular practice. Infections and complications are common, morbidity, and mortality continue. In contrast, some patients with congenital transposition of the great arteries (c-TGA) have undergone surgery in childhood, while others appear for the first time in adulthood, and the diagnosis is often missed (23).

Conclusion

Transposition of the Great Arteries (TGA) causes the aorta and right ventricle, which are "transposition of the great arteries," to reverse from the original heart. Unlike the normal anatomy of the heart, the aorta connects to the right ventricle and the pulmonary artery connects to the left ventricle. TGA is a universal term, also known as Dextro-transposition of the great arteries (D-TGA) and Levo-transposition of the great arteries (L-TGA), and also known as congenitally corrected transposition of the great arteries. When these arteries are distorted, oxygen-poor (blue) blood flows from the body into the right atrium, right ventricle,

aorta, and body. Oxygenated (red) blood, unlike normal blood circulation, returns from the lungs to the left atrium, enters the left ventricle and is sent back to the lungs.

Most babies with TGA are born with a small hole in the middle of their atria that allows enough red blood to enter the body to sustain life for a few hours. TGA is usually diagnosed within the first few hours after birth and is life-threatening, and infants desperately need specialized therapy to survive. The most common initial treatment used is atrial balloon septostomy, which uses a balloon on the end of a catheter (a small flexible tube) to enlarge the opening between the atria. Complete open heart repair usually occurs a few days later. If there are no unusual risk factors, more than 98 percent of operated babies survive infancy. Most children who have TGA surgery recover and grow normally, although they may be at risk for arrhythmias, leaking valves, and other heart problems in the future.

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