Transposition of the Great Arteries: Diagnosis, and Treatment

Note: The name of the author should be written as (Omar Elsaka), don't write it as (Omar Mohamed Raouf Mohamed Ahmed Elsaka). please, just write it as (Omar Elsaka).

Abstract

Background: The cyanotic congenital cardiac disorder known as transposition of the great arteries (TGA) affects neonates. Ventriculoarterial dissonance, in which the aorta emerges from the anatomically right ventricle and the pulmonary artery emerges from the morphologically left ventricle, is a characteristic aspect of Transposition of the Great Arteries. The anatomy of the coronary artery is abnormal in patients with major arterial transplantation: the most common types are the left coronary artery circling the left coronary artery and exiting the right coronary artery (22 percent), single right coronary artery (9.5 percent), left coronary artery (3 percent), and reverse origin of the coronary arteries (3 percent).

Conclusion: If there are no particular risk factors, more than 98 percent of surgically treated newborns survive childhood. ~~More than 98 percent of surgically treated infants survive childhood if there are no unique risk factors.~~ Most children who have TGA surgery recover and grow normally, while they may be at low risk for arrhythmias, leaky valves, and other cardiac problems in the future.

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

Introduction

The two main arteries from the heart are reversed in transposition of the great arteries, a dangerous but uncommon cardiac abnormality that occurs at birth (congenital). Dextro-transposition of the major arteries is another name for this disorder. Levo-transposition of big arteries is a rare variation of this disease. Large artery dislocation is frequently identified in the weeks leading up to birth or in the early hours of life. The most common treatment is corrective surgery after birth. It's hazardous to have a kid with transposition of the great arteries, but with the appropriate treatment, the future looks bright (1).

Causes and Risk Factors

Low-oxygen blood circulates in the right side of the heart, bypassing the lungs and returning to the body. The left side of the heart circulates oxygen-rich blood, which returns to the lungs

without circulating to other regions of the body. The skin develops cyanosis when low-oxygen blood circulates throughout the body. Transposition of big arteries is referred to as cyanotic congenital heart disease because of this. Genetics, rubella, other viral infections in pregnancy, diabetes in moms over 40 years old, or their mothers may all raise the incidence of this disorder, but the cause is usually unclear (2).

In adults, the right ventricular might eventually stop functioning as the main pump chamber, resulting in heart failure. The tricuspid valve, which protects the right ventricle in adults, is frequently dysfunctional. Heart failure can be caused by valve and ventricular dysfunction. Valve replacement, ventricular support devices, and heart transplants may be required in the future. Risk factors include: The specific origin of aortic blockage is unknown, however various variables, such as rubella or another maternal viral disease with a history of pregnancy, may raise the likelihood of giving birth to a baby with this condition. Consumption of alcoholic beverages during pregnancy, smoking throughout pregnancy, and moms with diabetes under control (3).

Pathophysiology of TGA

The systemic and pulmonary circulations are opposed. The systemic venous blood is pumped to the lungs in a systemic circulation without oxygen after returning to the right heart. Instead of traveling to the rest of the body, oxygenated blood from the left heart returns to the lungs. Unless unsaturated and oxygenated blood combines in one or more pathways, this disparity is incompatible with life (e.g., an arterial, ventricular, or large arterial pathway) (4).

Types of TGA

A cyanotic heart condition in which the aorta starts in the right ventricle and the pulmonary artery originates in the left ventricle is known as dextro-transposition of the major arteries of the aorta. This switch sends unoxygenated blood from the right heart straight into the aorta, bypassing the lungs and coursing throughout the body and heart. The left heart does not return to the body as usual in this state, instead of sending oxygen-rich blood to the lungs via the pulmonary arteries. As a result, there are two "parallel" systems. Because of a lack of oxygen, the newborn turns blue (cyanosis). This is known as cyanotic congenital heart disease (CAD) (figure 1) (5).

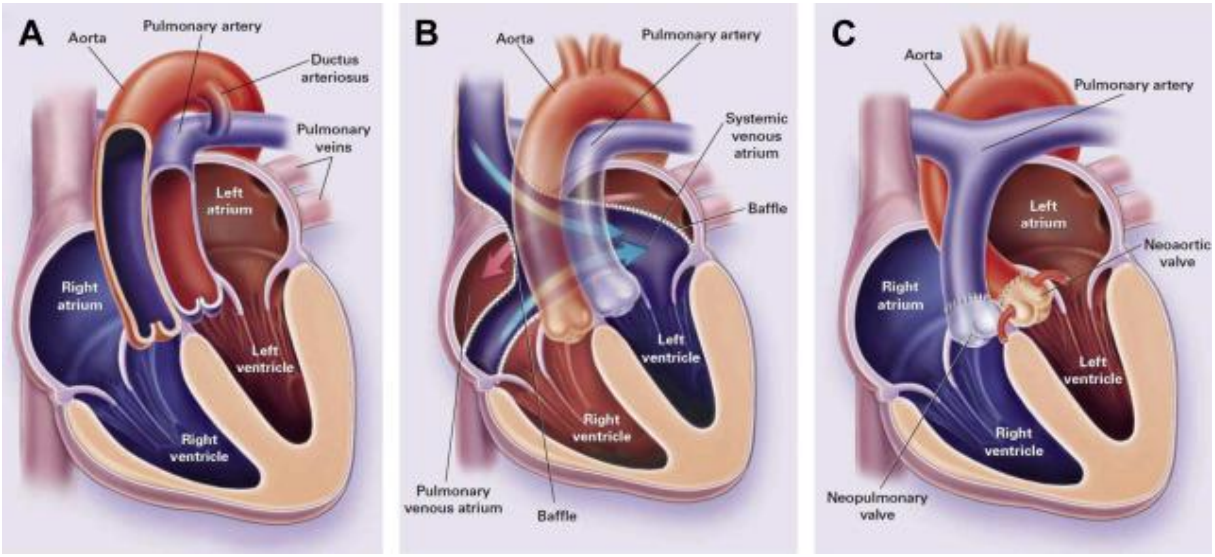

Figure 1 Dextro-Transposition of the Great Arteries (5)

The main arteries shift to the left of the anterior aorta in Levo-transposition of the great arteries, an uncommon cardiac condition. The pulmonary artery, as well as the left and right ventricles, move in tandem with their atrioventricular valves. To put it another way, the right ventricle is on the left side of the heart, while the left ventricle is on the right. Low systemic blood flow is a symptom of this illness. Despite its great resistance to the systemic circulation, the right ventricle, which prefers low-pressure pumping of blood to the pulmonary circulation, can cause difficulties owing to pressure shifts since it must pump blood at very high pressures. Because now it is in the position in which the left ventricle normally is (figure 2)(6).

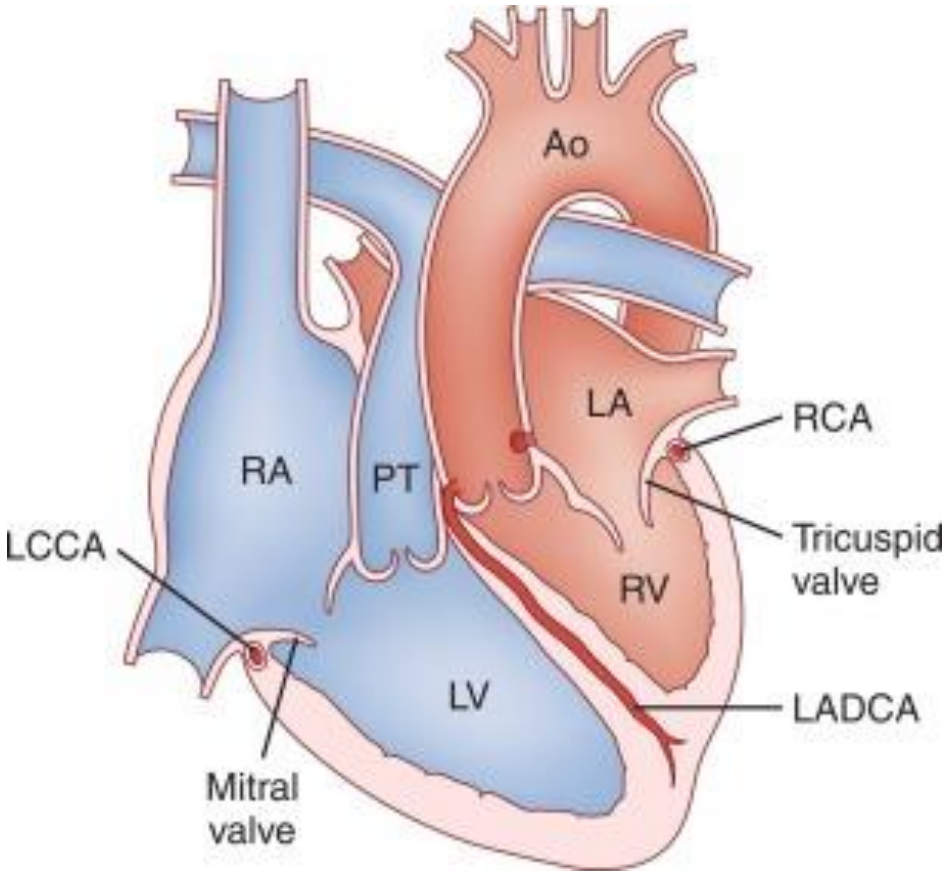

Figure 2 Levo-Transposition of the great arteries (6)

Simple and complex TGV: TGV is frequently linked with various heart abnormalities, with the patent foramen ovale, ventricular septal defect, and patent ductus arteriosus is the most common intracardiac shunts. There may be constriction or other problems in the valves and/or blood arteries in people with an atrial septal defect. TGV is referred to as "simple" if there are no other cardiac abnormalities. TGV is said to as "complicated" if there are additional flaws (7).

Complications

Oxygen deprivation in tissues is a possible consequence of a big artery transposition: infant tissues receive insufficient oxygen (hypoxia). Your infant will not survive unless he has a blend of oxygenated and non-oxygenated blood. A heart attack occurs when the heart is unable to pump enough blood to meet the body's needs, and the right ventricle pumps at a higher pressure than usual. The condition worsens with time. The right ventricular muscles may harden or weaken as a result of the increased pressure. Shortness of breath is caused by lung damage. All infants with big artery implants require surgery early in life, usually within the first week. The operation to rectify the transposition when it is discovered in youngsters is the most prevalent type of surgical arterial switch. The surgeon adjusts the big arteries so that they are appropriately attached to the pumping chamber during this procedure. The coronary arteries, which provide blood to the heart, must also be replaced. Although this procedure saves lives, it can lead to complications later in life, such as narrowing of the arteries that supply blood to the heart (coronary arteries), abnormal heart rhythms (arrhythmia),

myocardial infarction (which can lead to heart failure), and narrow vessels connecting large vessels and leaky heart valves (8).

Symptoms

Within hours of birth, severe cyanosis sets in, followed by metabolic acidosis caused by inadequate tissue oxygenation. Symptoms and signs of heart failure (e.g., tachypnea, dyspnea, tachycardia, sweating, inability to gain weight) can develop in the first few weeks of life in patients with a moderate or severe atrial septal defect, a ventricular septal defect, a patent ductus arteriosus, or a combination of these conditions. The physical exam is unimportant except for extensive cyanosis. There can't be a cardiac murmur unless it's accompanied by abnormalities. The second heartbeat (S2) is quite 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. If a baby's skin turns blue or he or she has trouble breathing after birth, doctors fear a cardiac condition, such as a TGA. If a child has another cardiac problem, such as a blue skin tone, it may be undetectable. Atrial septal defect refers to a hole in the upper chamber of the heart. A ventricular septal defect is a condition that affects the lower chambers of the heart. It's also possible that the child's ductus arteriosus is patent. It is the junction of the aorta and the pulmonary artery, the two largest blood channels in the heart, where oxygen-rich blood and unoxygenated blood can mix (10).

As your baby becomes more active, heart problems prevent enough blood from flowing, resulting in cyanosis. If the child's doctor detects a heartbeat, which is an unnatural wheezing sound caused by poor blood flow, he may suspect heart failure. A physical examination is insufficient to diagnose major artery transposition properly. One or more of the tests listed below are required for accurate diagnosis (10).

Echocardiogram

An echocardiogram is a type of heart ultrasound study in which sound waves from your baby's heart are converted into moving images that may be viewed on a video screen. Based on the state of the aortic and pulmonary arteries, doctors employ this test to detect major artery translocations. A heart defect such as ventricular dysfunction, atrial septal defect, or ductus arteriosus may be shown by echocardiography(figure 3) (11).

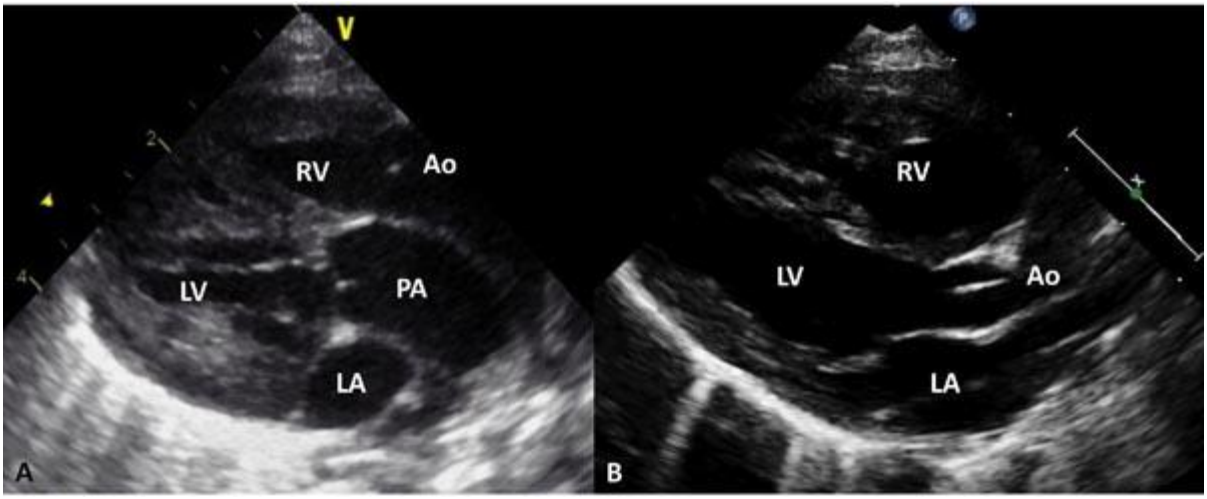

Figure 3 Echocardiogram of Transposition of the Great Arteries (A), Normal (B) (11)

Cardiac catheterization

When other tests, such as echocardiography, do not provide enough information for a diagnosis, this procedure is frequently used. A thin, flexible tube (catheter) is inserted into the artery or vein of the baby's groin and carried to his or her heart during cardiac catheterization. The dye is administered through a catheter to allow an X-ray to be taken of the baby's heart architecture. The catheter can assess the amount of oxygen in the blood as well as the pressure in the heart chambers and blood arteries. The temporal displacement of the immediate major arteries can be treated with cardiac catheterization (balloon atrial septostomy) (figure 4) (12).

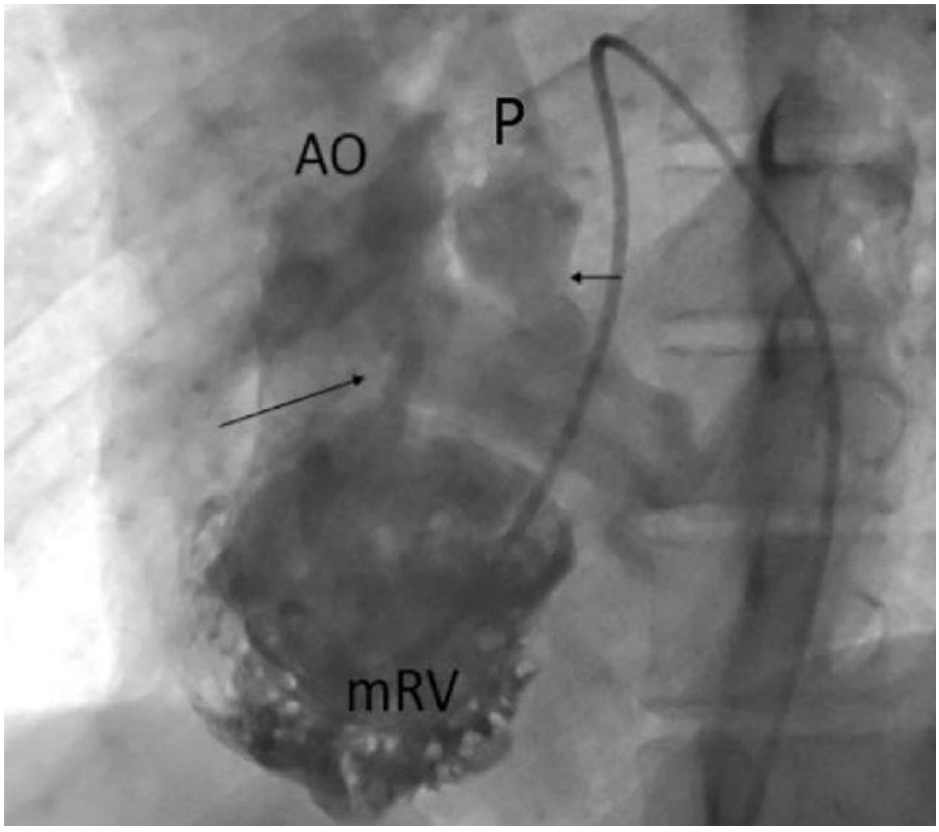

Figure 4 cardiac catheterization of a CCTGA and Situs Inversus patient showed the anteriorly placed aorta arising from the morphologic right ventricle and the posteriorly placed pulmonary artery arising from the morphologic left ventricle and in association with ventricular septal defect (long arrow),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

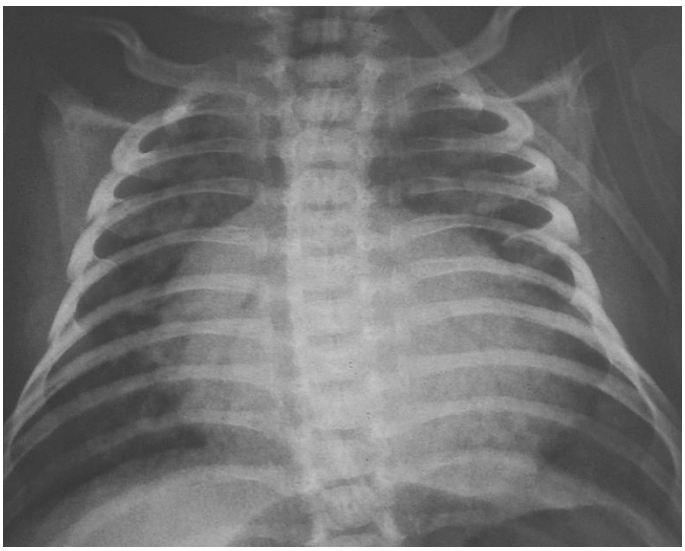

Figure 5 Chest X-ray of Transposition of the Great Arteries (13)

Electrocardiogram

With each heartbeat, an electrocardiogram captures the electrical activity of the heart. Patches with wires (electrodes) are attached to the baby's chest, wrists, and ankles during this procedure. Electrical activity is measured with electrodes and recorded on paper (Figure 6). (14). ~~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).~~

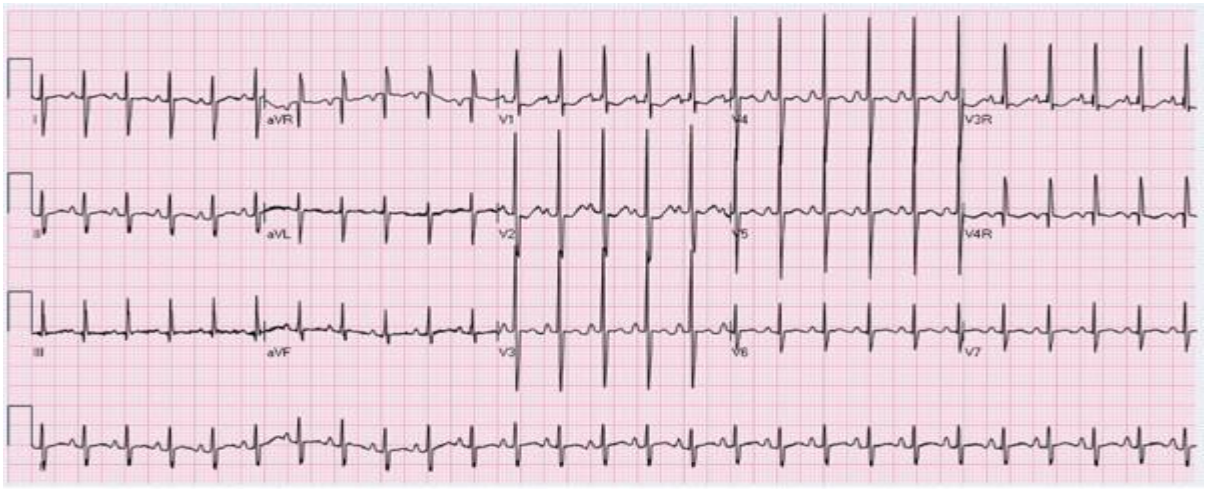

Figure 6 Electrocardiogram of Transposition of the Great Arteries (15)

Treatment

Before surgery

Before corrective surgery, a cardiologist can recommend several methods to assist control the disease. Drug: Prostaglandin E1 promotes blood circulation and oxygen-rich blood mixing by keeping the link between the aorta and pulmonary artery (ductus arteriosus) open. Atrial septostomy: This procedure, which improves the natural connection between the upper chambers of the heart, is usually performed by cardiac catheterization rather than surgery. This permits oxygen-rich and oxygen-depleted blood to mix, resulting in an improved oxygen supply to the baby's body (16).

Surgery

The surgical procedure is determined by the patient's age at the time of presentation, the presence of accompanying congenital heart defects, and the cardiothoracic surgeon's experience with a particular surgical technique. The majority of term infants with simple TGA can have their arteries switched in a minimally invasive procedure (figure 7) (17).

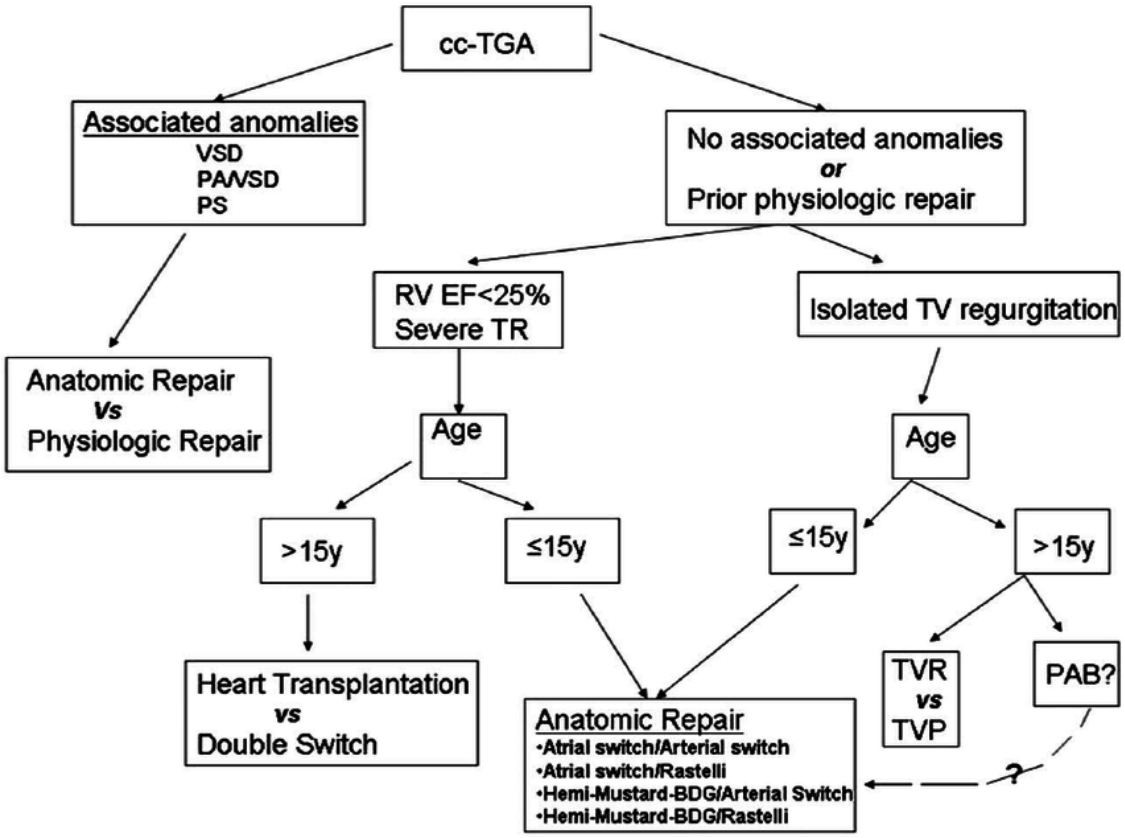

Figure 7 Decision making in choosing the type of surgery, cc-TGA: congenitally corrected transposition of great arteries, BDG: bidirectional Glenn, EF: ejection fraction, PA: pulmonary artery, PAB: pulmonary artery banding, PS: pulmonary stenosis. RV: right ventricle, TR: tricuspid regurgitation, TV: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

Transposition of the great arteries with intact ventricular septum

Arteriovenous switch surgery is the best option. It establishes ventriculoatrial compliance and represents anatomical progress. Because the left ventricle may not be able to sustain the systemic pressure in the low-pressure, low-resistance pulmonary circuit for long periods, this treatment should be performed when the newborn is less than 4 weeks old. Dissection of the coronary artery may not be possible in some cases, depending on the specific anatomy of the coronary artery (eg, intramural coronary artery), and arterial switching is not recommended. Changes in atrial levels appear to minimize short-term and post-surgical morbidity and mortality in this cohort (figure 8) (19).

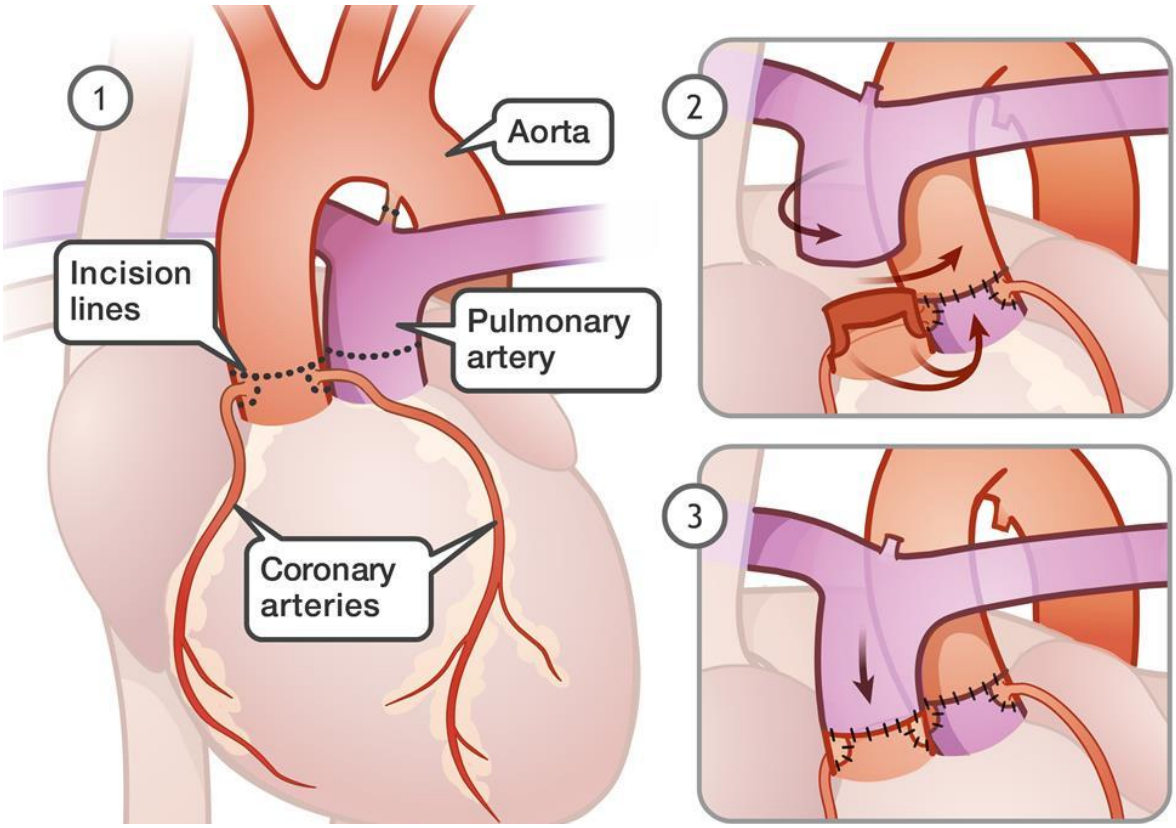

Figure 8 Transposition of the great arteries with intact ventricular septum (19)

Transposition of the great arteries with ventricular septal defect

The arterial switch surgery with interventricular septal closure is the favored method. A rastelli-type intracardiac repair is an option if the ventricular septal defect is extensive and complex, and the structure of the coronary artery makes the artery swap surgery ineffective. It may be advisable to wait until the child is older to perform a Rastelli-type procedure, as the right ventricular pulmonary artery must be severed during Rastelli surgery. If a kid develops severe congestive heart failure (along with growth retardation), reconstructive surgery or, if that is not possible, methods to restrict pulmonary blood flow may be recommended (figure 9)(20).

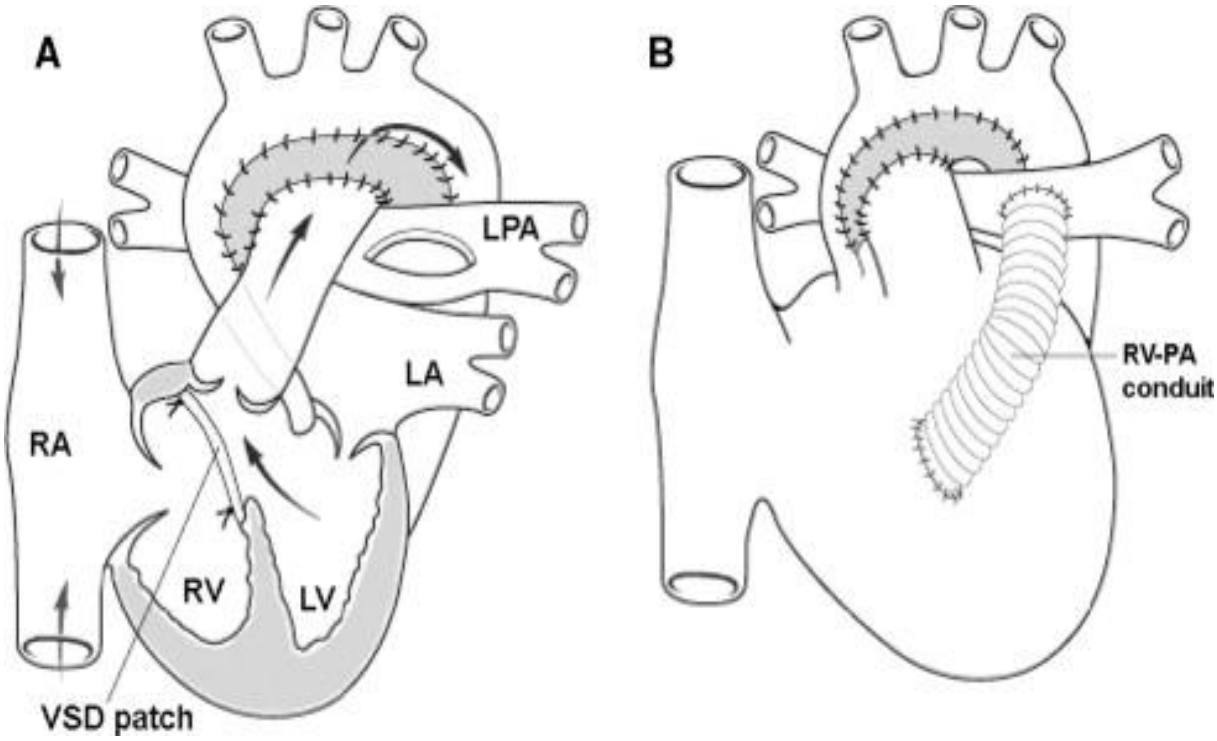

Figure 9 Rastelli procedure (20)

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

An arterial switch procedure can be hampered by stenosis or occlusion of the pulmonary valve (left ventricular outflow tract). If the ventricular septal defect is not confined and removed from the aorta, Rastelli intracardiac repair is conceivable. Because the Rastelli operation necessitates the creation of a route from the right ventricle to the pulmonary artery, it may be best to wait until the child is older to recuperate. In this instance, an aortic-pulmonary artery bypass may be required throughout the neonatal period to ensure enough 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

The surgeon constructs a tunnel (septum) between the two upper chambers of the heart during this procedure (atria). Deoxygenated blood is sent to the left ventricle and pulmonary artery, whereas oxygen-rich blood is directed to the right ventricle and aorta. In this surgery, the right ventricle, like a normal heart, is responsible for pumping blood not only to the lungs but also to the rest of the body. Arrhythmias, septum obstruction or leakage, and heart failure due to

long-term issues with right ventricular function are all possible side effects of atrial switch surgery (figure 10)(21).

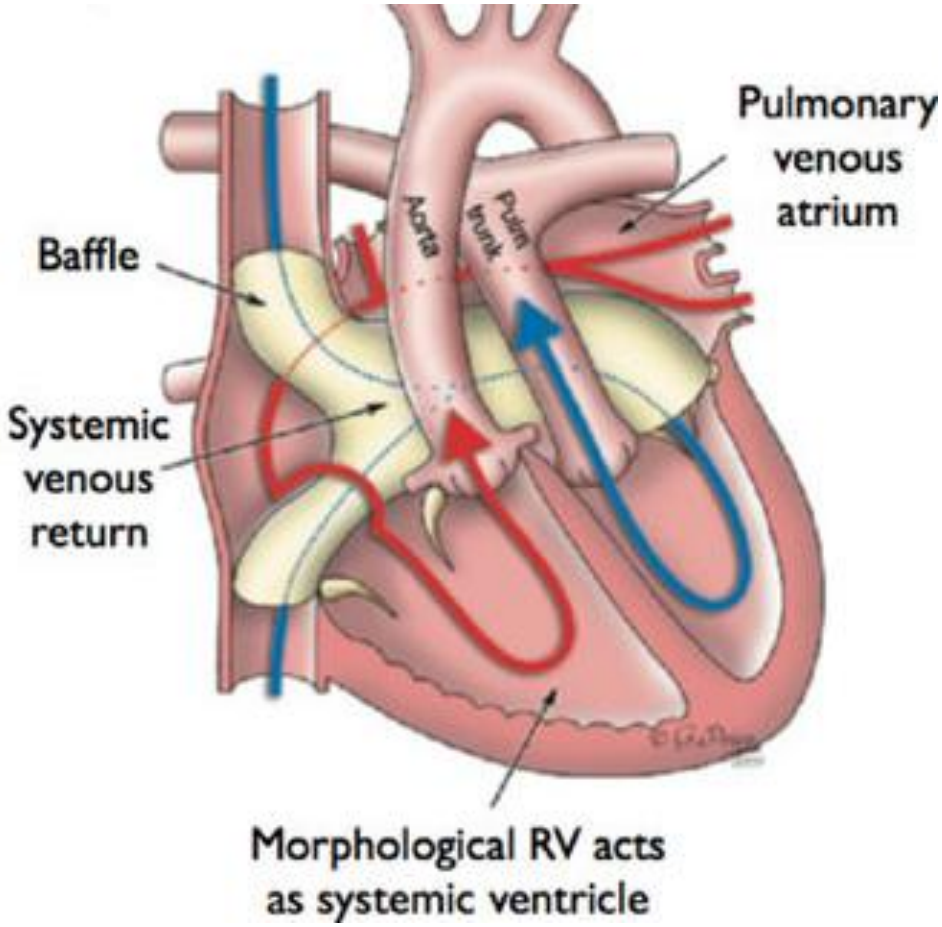

Figure 10 atrial switch operation (Mustard/Senning) for transposition of the great arteries. Systemic (blue) blood is directed from the superior caval vein and inferior caval vein into the left atrium, then via the mitral valve to the left ventricle and then to the pulmonary artery. Pulmonary (red blood) 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 newborn will require lifelong follow-up care from a cardiologist who specialises in congenital heart disease to assess their heart health. ~~The baby will need lifelong follow-up care from a cardiologist who specializes in congenital heart disease to evaluate their heart health after corrective surgery.~~ Cardiologists may advise the youngster to avoid activities like weightlifting or competitive sports, which can elevate blood pressure and put a strain on the heart. Many people who have artery swap surgery may not require any further procedures. Some consequences, such as arrhythmias, heart valve leakage, or heart-pumping issues, may necessitate further therapy (22).

Long-term care

The infant will need to be monitored and treated for the rest of his life. The youngster will be monitored and follow-up visits with a pediatrician will be scheduled regularly. As the infant grows older, he or she will be looked after by a congenital heart expert who will be able to track their progress. A big artery transplant can have long-term consequences for a child's adult life, as it can cause various health issues. Adults with congenital cardiac abnormalities may require additional care (22).

Pregnancy

A safe pregnancy is feasible if the transposition of the great arteries is corrected in childhood, but it may necessitate specialized treatment. Consult a cardiologist or obstetrician before conceiving if the patient intends to become pregnant. Pregnancy can put the patient and her baby in danger if she develops issues like arrhythmias and significant cardiac abnormalities. Even with transposition repair, pregnancy is not indicated in some cases, such as in women who have major difficulties owing to a cardiac condition (22).

Discussion

Many adolescent patients with ventricular arterial incompatibility made it to adulthood. Arterial switching (mustard or sensing surgery) is a procedure in which the anatomic right ventricle (RV) supports systemic circulation in people who have complete transposition of major arteries. Regurgitation of the RV and tricuspid valves is very prevalent. Some patients may require a heart transplant in the future. With longer follow-up, sinus node dysfunction becomes more common, and some patients require a pacemaker. Arterial rhythm is a common occurrence. Patients who undergo arterial switching also make it through adolescence. Coronary artery stenosis, pulmonary artery dysfunction, and aortic regurgitation are all long-term issues. Atrioventricular and ventricular arterial deterioration are present in patients with congenital transplant correction, resulting in morphological RV and thin tricuspid valves in the systemic circulation. Associated defects such as tricuspid valve abnormalities, ventricular septal defects, and pulmonary stenosis affect the majority of patients. Heart block develops as people get older (23).

Atrial arrhythmias are common, and their existence should make it easier to investigate hemodynamic issues. Progressive tricuspid regurgitation develops as people become older, and it's linked to poor right ventricular function. At the first symptom of right ventricular dilatation or malfunction, surgical therapy should be explored. All patients who specialize in the clinical, radiological, and hemodynamic evaluation of congenital heart disease in adults should make regular visits to the center. Many people who had the transposition complex lived to be adults. Complete transposition patients have been exposed to earlier repairs and are increasingly being seen in cardiovascular practice. Infections and complications are common, and mortality and morbidity are still high. 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

The aorta and right ventricle, which are "transposition of the great arteries," reverse from the original heart in Transposition of the Great Arteries (TGA). The aorta connects to the right ventricle, whereas the pulmonary artery connects to the left ventricle, in contrast to normal heart structure. TGA stands for transposition of the great arteries, and it is also known as Dextro-transposition of the great arteries (D-TGA) and Levo-transposition of the great arteries (L-TGA), as well as congenitally repaired transposition of the great arteries. Oxygen-poor (blue) blood flows from the body into the right atrium, right ventricle, aorta, and body when these arteries are twisted. 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 TGA babies are born with a small hole in the middle of their atria, allowing enough red blood to enter the body to keep them alive for a few hours. TGA is a life-threatening condition that is usually discovered within the first few hours of delivery. Infants who survive TGA require specific treatment. The most frequent initial treatment is atrial balloon septostomy, which enlarges the opening between the atria using a balloon on the end of a catheter (a short flexible tube). A few days later, the open heart is repaired completely. More than 98 percent of operated babies survive infancy if there are no unique risk concerns. 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.

Conflict of interest

It is not applicable

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