

Transposition of the Great Arteries

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Transposition of the Great Arteries:

Introduction:

Transposition of the great arteries (TGA) is the most common cyanotic congenital heart defect (CHD) in the neonate. It constitutes 5% of all CHDs and 10% of all neonatal cyanotic CHDs¹. A number of definitions have been used to describe TGA, but the most accurate description is “a defect in which the aorta arises from the morphologic right ventricle and the pulmonary artery from the morphologic left ventricle” (Figure 1). Therefore, the circulation is parallel instead of normal in-series circulation. Therefore, the pulmonary venous blood does not get delivered to the body and the systemic venous blood does not get oxygenated. Infants will not live unless there are inter-circulatory connections such as atrial or ventricular septal defect or a patent ductus arteriosus.

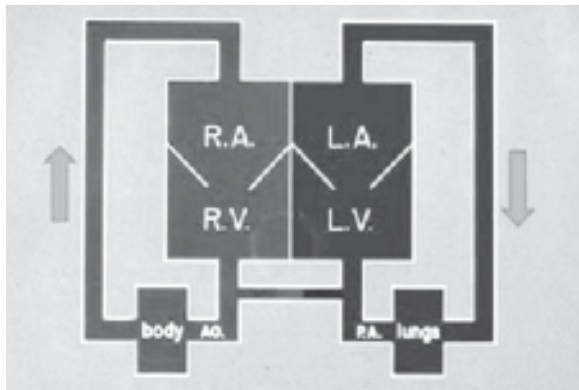


Fig-1: Box diagram of the heart showing parallel circulations in transposition of the great arteries.

Classification:

The TGA patients are arbitrarily divided into: Group I, TGA with intact ventricular septum; Group II, TGA with ventricular septal defect (VSD), and Group III, TGA with VSD and pulmonary stenosis (PS).

Associated Defects:

1) Malaligned outlaid VSD: as it is related to the uppermost portion of tricuspid annulus, tricuspid valve abnormalities are seen such as straddling TV with chordal attachment to the LV, Overriding of tricuspid annulus, TV tissue

protruding through VSD and causing subpulmonary obstruction.

2) Subaortic VSD: In these patients the aorta is usually L-posed (Left and anterior).

3) Anterior displacement of infundibular septum is associated with overriding of pulmonary annulus to the RV and severe forms will be hemodynamically similar to Taussig-Bing type DORV. Anterior displacement of infundibular septum causes subaortic stenosis, is frequently associated with aortic arch hypoplasia, coarctation or complete interruption of the aortic arch.

4) Posterior malalignment of the infundibular septum is associated with varying degrees of left ventricular outflow tract obstruction - subpulmonary stenosis, annular hypoplasia or even pulmonary valvar atresia. In about 5% of the patients, left ventricular outflow tract (LVOT) obstruction (or subpulmonary stenosis) occurs. The obstruction may be dynamic or fixed. Dynamic obstruction of the LVOT, which occurs in about 20% of such patients, results from bowing of the interventricular septum to the left because of a high RV pressure. Anatomic (or fixed) subpulmonary stenosis or abnormal mitral chordal attachment rarely causes left ventricular outflow tract obstruction.

Clinical Features

Clinical features depend upon the anatomic type.

Symptoms In Group I with intact septum, infants usually present with cyanosis within the first week of life (sometimes within hours to days of life). They may otherwise be asymptomatic. However, they will, with time, develop tachypnea and respiratory distress. If they are not appropriately treated, they become acidotic and go on to become lethargic without lack of spontaneous movement, and eventually die. Group II TGA patients with VSD present with symptoms of congestive heart failure (tachypnea, tachycardia, sweating, and poor feeding) between 4 to 8 weeks of life, but the cyanosis is minimal. Group III patients (TGA with VSD and PS) have variable presentation, depending upon the severity of PS and the

degree of inter-circulatory mixing. If there is poor mixing, they may present early in life and mimic TGA with intact septum. If the PS is severe, the presentation is essentially similar to that seen with Tetralogy of Fallot (TOF)². With moderate PS the presentation is late with longer survival. With mild PS, congestive heart failure signs may be present, similar to Group II patients.

Physical Examination

The Group I patients with intact septum usually have severe cyanosis, but are without distress until severe hypoxemia and acidosis develop. Clubbing is not present in the newborn period and may not develop until 3 to 6 months. The right ventricular impulse is increased and the second heart sound is single. No cardiac murmurs are present; occasionally a grade I-II/VI nonspecific ejection systolic murmur may be heard along the left sternal border.

In Group II patients, tachypnea, tachycardia, minimal cyanosis, hepatomegaly, increased right and left ventricular impulses, single second sound, a grade III-IV/VI pansystolic murmur at the left lower sternal border and a mid-diastolic flow rumble (murmur) at the apex may be present.

In Group III patients, the findings are similar to TGA with intact septum, TGA with VSD, or TOF depending upon the degree of mixing and severity of PS. Most of them however, will have a long ejection systolic murmur at the left upper sternal border and/or a pansystolic murmur at the left lower sternal border; both murmurs are usually grade III to IV/VI in intensity.

Noninvasive Evaluation:

Chest X-ray

In Group I patients with intact ventricular septum, chest roentgenogram looks benign with normal to minimal cardiomegaly and normal to slightly increased pulmonary vascular markings (Figure 2). The shadow of the thymus rapidly involutes and a narrow pedicle (superior mediastinum) may be seen. A combination of the above signs may sometimes appear as an "egg-shaped" heart on a postero-anterior chest film. In Group II patients with VSD, moderate to severe cardiomegaly and increased pulmonary vascular markings are usually seen. In Group III patients, mild to, at worst moderate cardiomegaly may be observed. The pulmonary vascular marking may be increased, normal or decreased, dependent upon the severity of PS.

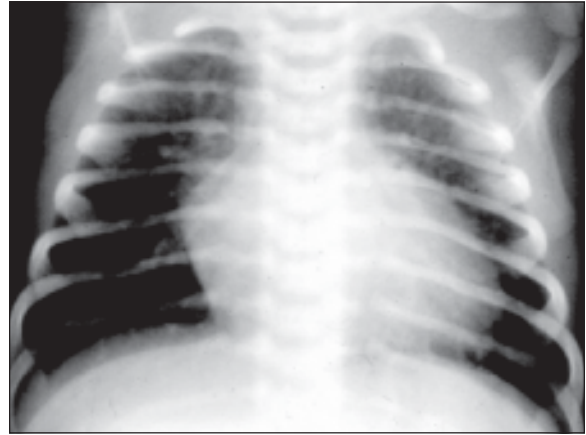


Fig-2: Chest radiograph of a two-day old infant with transposition of the great arteries demonstrating mildly enlarged size of the heart and increased pulmonary vascular markings

Electrocardiogram

The electrocardiogram in a neonate with TGA and intact septum (Group I) may be normal with the usual right ventricular preponderance seen at this age. In older infants clear-cut right ventricular hypertrophy becomes obvious and, in addition, right atrial enlargement may be seen. In Group II patients, biventricular hypertrophy and left atrial enlargement are usual. In Group III, right ventricular or biventricular hypertrophy is seen.

Echocardiogram

Echocardiogram is helpful in the diagnosis and assessment. A helpful indirect sign is a somewhat posterior of the great vessel arising from the left ventricle in a precordial long axis view, indicating the vessel is pulmonary artery in contradistinction to anteriorly coursing ascending aorta. The presence of an interatrial communication and patent ductus arteriosus and shunt across them by color and pulsed Doppler should also be evaluated. In addition to these, demonstration of VSD and PS will place the patients into the respective groups.

Other Laboratory

Blood gas values are useful in demonstrating the degree of hypoxemia and ventilatory status. Serum glucose & calcium level should also be monitored.

Cardiac Catheterization and Angiography

With the increased accuracy of echocardiographic diagnosis, invasive studies are not necessary for diagnosing TGA. Need for rapid relief hypoxemia and acidosis by balloon atrial septostomy and the need for a greater definition of coronary artery anatomy prior to

arterial switch procedure may necessitate catheterization and angiography. Selective right ventricular angiography (Figure 3) reveals a morphologically right ventricle with opacification of an anteriorly and superiorly displaced aorta. The aortic valve is located to the right of the pulmonary valve (d-TGA). The aorta ascends in a normal fashion and usually descends on the left side of the spine. If a VSD is present, it may be visualized. A laid-back view of the aortic root angiography along with a lateral view may be useful in demonstrating coronary artery anatomy. Left ventricular cineangiogram (Figure 3) reveals a morphologic left ventricle with prompt opacification of the pulmonary.

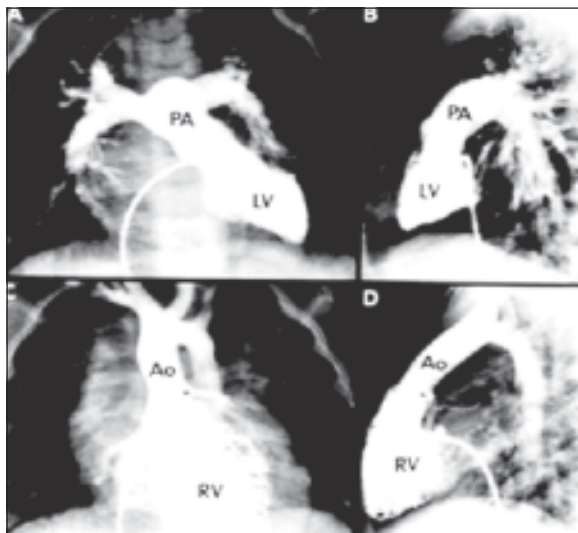


Fig-3: A & B. Left ventricular (LV) cineangiogram in posteroanterior (A) and lateral (B) views demonstrating a finely trabeculated, morphologic left ventricle with prompt opacification of the pulmonary artery (PA)

Management

The treatment of choice in the neonates with TGA is total surgical correction by arterial switch procedure (Jatene)¹⁰. However, since the surgery is usually performed at about the age of 7 days, the infant should be cared for to ensure good clinical and metabolic state before going to surgery.

General Measures

Initial management of TGA is similar to that used in other cyanotic neonates. The infant's temperature should be monitored and neutral thermal environment maintained. Ambient oxygen should be administered if the infant is hypoxic. Metabolic acidosis, defined as pH < 7.25 should be corrected with sodium bicarbonate. Since hypoglycemia can be a significant problem, the infant's serum glucose should be monitored.

Palliative Therapy

If untreated, TGA with intact septum carries a poor prognosis. Instead of having a normal in-series circulation, the TGA patients have parallel circulation (Figure 1). Without either an intra-cardiac or extra-cardiac shunt, the infants with TGA will not survive (Figure 4). The fetal circulatory pathways {patent foramen ovale (PFO) and patent ductus arteriosus (PDA)} will provide some mixing initially. However, in most neonates with TGA, the PFO and PDA tend to undergo spontaneous closure and the infant gets progressively hypoxic. The PDA and PFO can be kept open or enlarged by pharmacological or mechanical means respectively.

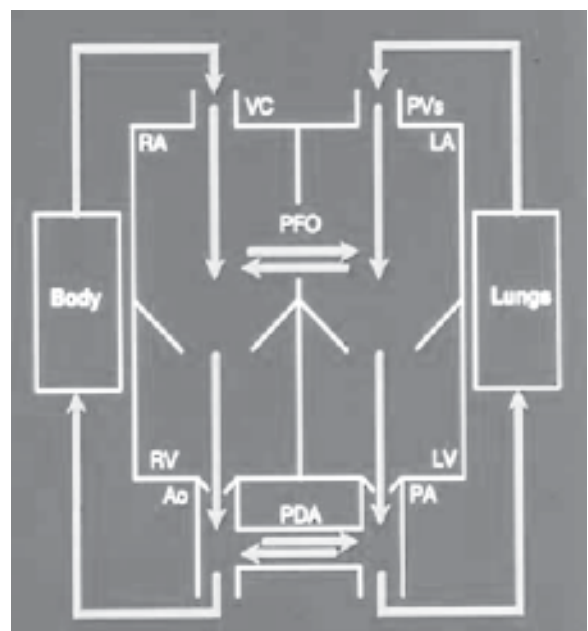


Fig-4: Box diagram of the heart showing parallel circulations in transposition of the great arteries.

Patent Ductus Arteriosus

Intravenous infusion of prostaglandin E₁ (PGE₁) (0.05 to 0.1 mcg/kg/min) may help open the ductus, thus improve oxygenation. If hypoxemia does not improve even after PGE₁, balloon atrial septostomy may become necessary.

Patent Foramen Ovale

Balloon atrial septostomy^{3,4} (Figure 6) has been extensively used in the palliation of neonates with TGA with intact septum. If naturally present PFO and/or PGE₁ infusion to dilate the PDA do not maintain reasonably good oxygen saturations (60 to 70% without metabolic acidosis), balloon atrial septostomy should be performed, preparatory to arterial switch procedure.

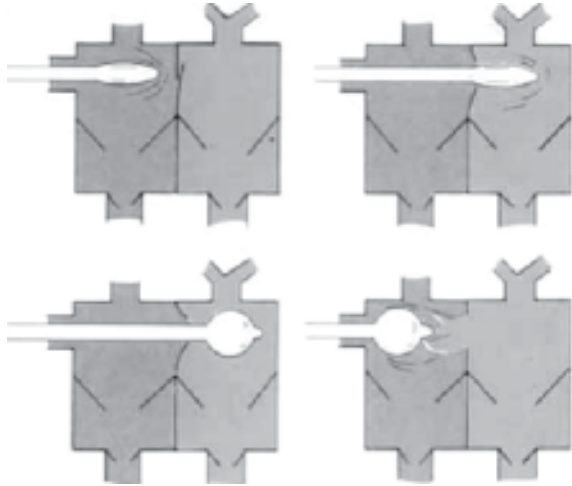


Fig-5: Diagrammatic display of the procedure of Rashkind balloon atrial septostomy. An un-inflated balloon septostomy catheter is placed in the right atrium (top left) and advanced across the patent foramen ovale into the left atrium (top right). The balloon is inflated with diluted contrast material (bottom left) and rapidly pulled back into the right atrium (bottom right), thus performing balloon atrial septostomy

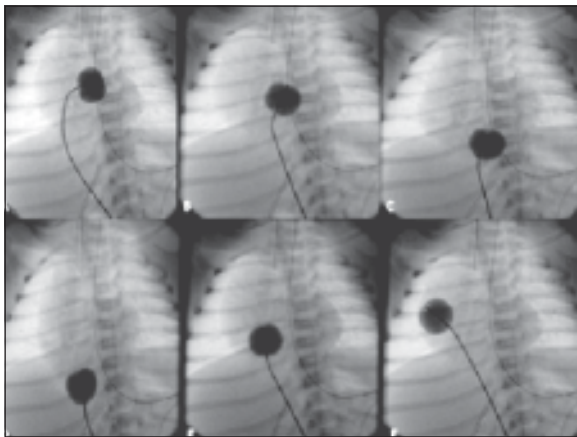


Fig-6: Selected cinefluoroscopic frames of the Rashkind's balloon septostomy procedure.

Septostomy Procedures

In 1966, Rashkind and Miller³ described a technique, now called Rashkind balloon atrial septostomy, which was extensively used to improve atrial mixing in neonates with TGA. The reason for success of balloon septostomy is a very thin and frail lower margin of the PFO (septum primum) in the newborn which can be torn by rapid withdrawal of an inflated balloon across the PFO. Some babies do have thick atria septa. To address these situations, Park and his

associates, in the mid/late 1970s, extended the utility of the balloon septostomy procedure by introducing blade atrial septostomy to enlarge defects with thick atrial septae.⁷ A built-in retractable blade (knife) cuts the lower margin of the patent foramen ovale (PFO) which is followed by balloon atrial septostomy. More recently, static balloon angioplasty,^{6,8,9} stents,¹⁰⁻¹² Brockenbrough atrial septal puncture,¹² radiofrequency ablation¹³⁻¹⁵ and cutting balloons were applied to create and/or enlarge the atrial defects⁴. In most patients conventional balloon atrial septostomy is all that is necessary to palliate TGA patients until surgery.

Rashkind Balloon Atrial Septostomy Procedure

In TGA patients who are stable, the hemodynamic (usually limited) data, including selective cine-angiography, as needed, are performed. If the infant is unstable or has extremely low oxygen saturations, one may proceed directly with balloon septostomy. In such situations, aortic saturation and pressure pullback across the atria and echocardiographic size of atrial defect are recorded. The balloon septostomy procedure involves inserting a balloon septostomy catheter, usually via a sheath percutaneously placed in the femoral vein, into the left atrium via the PFO. The balloon is inflated with diluted contrast material to a sub-maximal amount (usually 2 to 3 ml) and rapidly pulled back across the atrial septum (Figure 5 & 6) after ensuring that the catheter tip is located in the left atrium either by lateral fluoroscopy or by echocardiography. Once the catheter is pulled back to the inferior vena cava, the catheter should be rapidly advanced into the right atrium; all this is done as a single motion (Figure 6). The balloon should be deflated as the catheter is repositioned into the right atrium. This jerking motion of the contrast filled balloon catheter produces a tear in the lower margin of the PFO (septum primum) with resultant bidirectional shunt. Increase in systemic arterial oxygen saturation, disappearance of pressure gradient across the atrial septum and echographic increase in the size of the atrial defect with non-restrictive Doppler flow across the atrial septum are demonstrated in successful procedures. Some workers balloon-size the atrial defect both prior to and following balloon septostomy. In the initial description of balloon septostomy by Rashkind and Miller,⁸ the catheter was introduced into the femoral vein by cut-down. To avoid femoral venous cut-down, insertion of the catheter and performance of balloon septostomy via the umbilical vein¹⁶ has been advocated. When percutaneous technology became available, the balloon catheter was introduced via appropriate sized percutaneously inserted femoral venous

sheaths. Our first choice is to perform balloon septostomy via the umbilical venous route. Therefore, we encourage our neonatology colleagues to place an umbilical venous line early on, with its tip well into the right atrium, before the ductus venosus constricts. At the time of septostomy, this line is exchanged over a wire with an appropriate sized sheath. Initially Rashkind balloon septostomy catheters were used. Because the catheters were straight, sometimes making it difficult to advance the catheter into the left atrium, and because of the limited volume of fluid that these balloons would take, most cardiologists have switched to Edwards septostomy catheters (American Edwards Baxter, McGaw Park, IL). These catheters have a gentle curve at the tip, facilitating easy access into the left atrium and larger volume of fluid that can be injected into these balloons.

TGA with VSD patients usually present with heart failure and aggressive anti-congestive measures are indeed needed. Balloon atrial septostomy may help relieve pulmonary venous congestion and improve oxygenation. These patients will require Jatene procedure along with closure of the VSD.

TGA with VSD and PS patients may have varying types of presentation. If poor mixing is the reason for hypoxemia, balloon atrial septostomy is the treatment of choice. If the hypoxemia is secondary to markedly decreased pulmonary flow, a Blalock-Taussig type of shunt may be needed. Sometimes both transcatheter balloon atrial septostomy and balloon pulmonary valvuloplasty may be needed to improve hypoxemia. Most of these patients eventually require a Rastelli type of repair.

Surgical Correction

Two types of surgical approaches, namely atrial (venous) and arterial switch are available for use. In the venous switch procedure, the systemic venous flow is directed towards the mitral valve and the pulmonary venous flow towards the tricuspid valve by constructing an intra-atrial baffle after the removal of the atrial septum. Jatene et al¹⁰ described anatomical corrections for TGA in 1975; they switched the aorta and pulmonary artery with relocation of the coronary arteries to the neo-aortic root. However, arterial switch procedure must be performed in the early neonatal period prior to deconditioning of the left (pulmonary) ventricle. Group III TGA patients with VSD and PS most often require Rastelli type of surgery in which left ventricular blood flow is directed into the aorta with

the VSD closing patch and a valved conduit (usually an aortic homograft) is inserted to connect the right ventricle to the pulmonary artery.

Summary and Conclusions

Transposition of the great arteries is a congenital heart defect in which the aorta arises from the right ventricle, while the pulmonary artery comes off the left ventricle. It is the most common cyanotic CHD in the neonate. In this condition the systemic and pulmonary circulations are parallel instead of the normal circulation which is in series. This anomaly is classified into TGA with intact ventricular septum, VSD and VSD with PS. The intact ventricular septum patients present in the very early neonatal period while the other two may present with symptoms slightly later. Cyanosis is the major symptom in intact septum patients, while heart failure is the presenting symptom in patients with TGA and VSD. TGA with VSD and PS have a variable presentation. Murmurs are notably absent in intact septum babies while loud pansystolic or ejection systolic murmurs dominate in the other two groups. While the chest x-ray and ECG are helpful in the diagnosis, echocardiographic studies are confirmatory in the diagnosis and quantification of the associated defects. PGE₁ to open the ductus and/or balloon atrial septostomy to enlarge the PFO may sometimes be required for palliation. Corrective surgery by arterial switch (Jatene) procedure is necessary in TGA patients with intact septum and those with VSD whereas Rastelli procedure may be required for TGA patients with VSD and PS.

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