

TRANSPOSITION OF GREAT ARTERIES

Definitions and Classification of TGA

Morphological right and left ventricles refer to the anatomic characteristics of the chambers and not their positions.¹

Atrioventricular discordance Inappropriate connections of the morphological right atrium to the morphological left ventricle and morphological left atrium to right ventricle.

Ventriculoarterial discordance: The pulmonary artery arises from a morphological left ventricle, and the aorta arises from a morphological right ventricle.

Complete or d-transposition of great arteries (TGA) denotes that the aorta arises from the morphological right ventricle, and the pulmonary artery arises from the morphological left ventricle (ie, there is ventriculoarterial discordance).

In **congenitally “corrected” or l-TGA**, there are inappropriate connections of the morphological right atrium to the morphological left ventricle and morphological left atrium to right ventricle (atrioventricular discordance and ventriculoarterial discordance) (Figure 1).

Complete TGA (d-TGA)

Anatomy and Pathophysiology

d-TGA refers to the normal rightward (dextro) bend of the embryonic heart tube and indicates that the inflow portion of the right ventricle is to the right of the morphological left ventricle. The aorta arises from the morphological right ventricle, and the pulmonary artery arises from the morphological left ventricle. The aorta also tends to be on the right and anterior, and the great arteries are parallel rather than crossing as they do in the normal heart. Because the systemic and pulmonary circulations run in parallel, there has to be a communication between them such as VSD, ASD or PDA. Without intervention the mortality rate is 90% by six months of age.¹ Expediting surgical repair could be neuroprotective in newborns with transposition. Surgery beyond 2 weeks of age is associated with impaired brain growth and slower language development in infants.²

Epidemiology

d-TGA occurs accounts for 5% of all forms of congenital heart disease and is one of the most common cyanotic defects.

Clinical Problems and Therapy in Adults

Adult patients with TGA have survived due to previous repair. Their condition depends on the mode of operation performed. *Atrial arrhythmias* (a marker of sudden death), *sinus nodal dysfunction*, *pulmonary hypertension* and *atrial baffle obstruction* are problems encountered in adulthood.

Atrial switch

Atrial switch or atrial baffle procedures are the **Senning** (creation of an atrial baffle from autologous tissue to direct the venous return to the contralateral atrioventricular (AV) valve and ventricle) or **Mustard** (excision of the atrial septum and creation of the baffle with synthetic material) operations.³ These operations leave the morphological RV to support the systemic circulation and **RV failure** and **TR** are common. Perioperative mortality is 20%, and 60% of patients are alive after 30 years of follow-up.⁴ Patients with d-TGA and asystemic RV face a variable but slow progression to-ward end-stage heart failure. Age, atrialarrhythmia, pacemaker, biventricular enlargement, systolic dysfunction, and tricuspid regurgitation were all associatedwith the primary outcome. Independent 5-year predictors of primary outcome were prior ventricular arrhythmia, heart failure admission, complex anatomy, QRS duration>120 ms, and severe right ventricle dysfunction based on echocardiography.⁵ Patients should be monitored with annual ECG and periodic monitoring for arrhythmias.⁶ CTI-dependent flutter is the most common arrhythmia in patients with D-Transposition of the Great Arteries and atrial switch. Transbaffle approaches were often necessary for ablation.⁷ Recommendations are provided in Figure 2 and Tables 1-3.

Rastelli operation

When d-TGA coexists with a large subaortic VSD and PS, the Rastelli procedure may be used. A patch is placed to direct flow from the LV to the aorta through the VSD, the PV is oversewn and continuity between RV and PV is established through a valve conduit. This operation has the advantage that the LV supports systemic circulation but **conduit degeneration** and **atrial arrhythmias** are common and **sudden death** may occur.^{6,8} Recommendations are provided in Table 4.

Arterial switch

The modern surgical approach is arterial switch that restores normal anatomy of circulation. This operation has replaced atrial switch procedures for D-TGA, and 90% of patients now reach adulthood.⁹ Arterial switch involves transection of the great arteries above the sinuses and restoration of their anatomic sites (ie the aorta to LV outflow tract if normal, and the pulmonary artery anterior to the aorta to the morphological right ventricle. The operation is performed in the first weeks of life or later as a two-stage procedure with PA banding, in the absence of PV stenosis, to “train” the LV in higher pressures. Early operation (3 days of life) is probably beneficial.⁹ Late arterial switch after “training” of the LV with a PV band can also be performed in young patients with previous atrial switch operations and failing RV. This strategy has the theoretical advantage of relieving the hemodynamic burden on the RV and tricuspid valve, potentially improving surgical results and longevity. Long-term and arrhythmia-free survival is excellent after arterial switch operation, and most patients maintain normal systolic function and exercise capacity.¹⁰ Complications include **chronotropic incompetence**, and **dilatation of the neo-aortic root with AR, intratrial re-entry tachycardia, branch pulmonary stenosis, PR**, and **coronary stenoses**. Obstructed coronary

arteries are seen in 5-7% of survivors and should be suspected in the presence of post-operative arrhythmias and ventricular dysfunction.¹⁰ Peri-operative mortality is 2.7% and rises to 5.3% in presence of a VSD.¹¹ Surgical repair of TGA performed in the developing world is associated with an early survival of 85%.¹² The patient after arterial switch operation represents a potentially higher long-term coronary risk, so it is reasonable to optimize CAD risk factors from young adulthood.⁶ The effect of ACE inhibitors on RV function is debated, but they may be indicated in symptomatic patients with RV dysfunction.¹³ Cardiac resynchronization therapy may be considered for the failing systemic RV.¹⁴ The AHA/ACC and ESC recommendations are presented in Tables 5 and 6.

Pregnancy

In patients with transposition of the great arteries, the risks associated with pregnancy are mainly attributable to women with a previous atrial (Senning and Mustard) switch. Women with an atrial switch operation tolerate pregnancy relatively well, but there is an increased risk of developing arrhythmias and RV failure.¹⁵ Women with a dilated neo-aorta require closer surveillance. The risk of congenital heart defect in the offspring is <5%. Endocarditis prophylaxis may be considered unless they have had an arterial switch procedure and have no residual valve dysfunction or outflow tract disturbance.

Congenitally Corrected Transposition (l-TGA)

Anatomy and Pathophysiology

There are inappropriate connections of the morphological right atrium to the morphological left ventricle and morphological left atrium to right ventricle. The right atrium enters the left ventricle, which gives rise to the pulmonary artery, and the left atrium enters the right ventricle, which gives rise to the aorta. The aorta is usually anterior and to the left, and the tricuspid valve always enters a morphological RV. Thus, the circulation continues in the appropriate direction but flows through the wrong ventricles. It is also called l-TGA since the morphological right ventricle is on the left of the morphological left ventricle.¹

Associated anomalies:

A **VSD** occurs in 70% of patients, usually in the perimembranous location.

Pulmonary stenosis occurs in 40% of patients and is commonly subvalvular either due to an aneurysm of the membranous septum or due to fibrous tissue or ring in the subvalvular area. Associated valvar pulmonary stenosis also occurs.

Abnormalities of the TV, especially inferior displacement resembling Ebstein's anomaly, occur in up to 70% of patients.

Due to the displacement of the AV node and His bundle **complete AV block** occurs at 2% per year. Tricuspid valve or VSD surgery may also precipitate heart block.

Epidemiology

l-TGA is a rare anomaly and comprises <1% of all forms of congenital heart disease.

Presentation

Patients may present for the first time in adulthood and the diagnosis is often overlooked. Some patients may be relatively normal from a functional standpoint, and survival to the eighth decade has been reported in the absence of associated anomalies. Failure of the systemic ventricle is much more common earlier in life, usually with concomitant tricuspid regurgitation.

Physical Findings

Physical signs are those of:

Systemic ventricular (RV) failure

Left AV valve regurgitation,

Complete AV block and SVT or AF

The cause of systemic ventricular failure is not established. Perfusion of the systemic ventricle by a single coronary artery (RCA) as well as systemic AV valve regurgitation are probably responsible.

Investigations

Chest Radiograph. With mesocardia or levocardia, the diagnosis may be suspected from the chest radiograph (Figure 3). The vascular pedicle appears abnormally straight because the normal arterial relationships are lost. The ascending aorta is not visible on the right side, and the convexities from the descending aortic knob and pulmonary artery are absent on the left side. l-TGA is one of the most common anomalies associated with dextrocardia and should be suspected when there is abdominal situs solitus (gastric bubble on the left) and dextrocardia (Figure 3).

ECG. The ECG may resemble inferior myocardial infarction with Q waves in the right precordial leads and absent Q waves in the left precordial leads. Various degrees of AV block may be present. Patients should be monitored with annual ECG and periodic monitoring for dysrhythmias.⁶

Echocardiography can be difficult, particularly with dextrocardia and mesocardia. Subcostal imaging facilitates detection of atrial situs and position of the cardiac apex and thus determines the presence or absence of dextrocardia or mesocardia. The morphological RV is on the patient's left (l-loop), and has prominent trabeculations. A high parasternal short-axis view may show the abnormal arterial relationships, with the aorta usually anterior and to the left of the pulmonary artery.

Cardiac magnetic resonance is now the imaging mode of choice (Table 7).

Electrophysiologic testing is indicated in patients with unexplained syncope and transposition of the great arteries with atrial switch surgery (PACES/HRS 2014 statement, I-C).¹⁶

Therapy

ACE inhibitors may be beneficial in symptomatic only patients with RV dysfunction.¹³

Surgical Repair

Various procedures have been used in adult patients. The physiologic strategy leaves the RV in a subaortic position and solely addresses associated anomalies, such as VSD or subpulmonary obstruction with or without an LV-to-pulmonary artery conduit, and has been associated with poor long-term survival related to systemic RV failure. In the more complex anatomic repair the morphologic LV is established as the systemic ventricle by performing an arterial switch (double switch operation) Rastelli/Nikaidoh procedure combined with an atrial switch (a Senning or a Mustard procedure) (Figure 3).¹⁷ Anatomic correction offers an up to 85% 10-year survival, and has superior results in patients with significant TR,¹⁸ resulting in higher overall and reintervention-free survival compared to physiologic repair.¹⁷ Systemic atrioventricular valve surgery remains a valuable option for the treatment of patients with ccTGA, with low early mortality and satisfactory long-term outcomes, particularly in those with systemic ventricular EF $\geq 40\%$.¹⁹ A double switch, ie a two-stage procedure with PA banding, in the absence of PV stenosis, to “train” the LV in higher pressures, is also used. A venous switch (either Mustard or Senning) and in those with normal LVOT arterial switch can be performed later. If a large VSD is present the Rastelli procedure may be used. Concomitant tricuspid valve surgery can also be performed if necessary. LV failure, AR and atrial arrhythmias are common problems with this approach the long-term results of which are unknown. Patients with corrected transposition should be scrutinized for the presence of systemic atrioventricular valve regurgitation. Valve replacement should be considered before systemic ventricular EF falls below 40% or the subpulmonary ventricular systolic pressure exceeds 50 mm Hg (Tables 8 and 9).^{8,20} In patients with a permanent pacemaker, regular echocardiographic monitoring is recommended because of the risk of worsening of systemic AV valve regurgitation with ventricular pacing (AHA 2015 statement, I-C).⁶ Cardiac resynchronization therapy may be considered for the failing systemic RV.¹⁴ In the presence of severe RV failure, transplantation is the only option.

Pregnancy

Systemic ventricular ejection fraction $<40\%$ or significant systemic AV valve regurgitation are contraindications for pregnancy (Table 10).²¹ Otherwise, pregnancy may be tolerated in most women with l-TGA but careful evaluation is mandatory. Endocarditis prophylaxis may be considered unless there is no valve dysfunction or outflow tract disturbance.

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