12

Transposition of Great Arteries & Arterial Switch

12.1 Morphology

The hallmark of transposition of the great arteries (TGA) is ventriculo-arterial discordance in which the aorta arises from the morphologic right ventricle (RV) and the pulmonary artery arises from the morphologic left ventricle (LV).

The anatomic subtypes of TGA are:

1. TGA with intact ventricular septum.
2. TGA with ventricular septal defect (VSD).
3. TGA with VSD and left ventricular outflow tract (LVOT) obstruction.
4. TGA with VSD and pulmonary vascular obstructive disease.

In two third of patients with TGA, the coronary artery anatomy is normal, but in 1/3 the coronary artery anatomy is abnormal, with a left circumflex coronary artery arising from the right coronary artery (22%), a single right coronary artery (9.5%), a single left coronary artery (3%), or inverted origin of the coronary arteries (3%).

12.2 Pathophysiology

The pulmonary and systemic circulations run in parallel rather than in series. Oxygenated pulmonary venous blood from the left atrium and left ventricle is recirculated to the pulmonary vascular bed via the pulmonary artery. Deoxygenated systemic venous blood from the right atrium and right ventricle is pumped into the systemic circulation, bypassing the lungs. This parallel circulation results in a deficient oxygen supply to the tissues and an excessive right and left ventricular workload. It is incompatible with survival unless mixing of oxygenated and deoxygenated blood occurs. The mixing of oxygenated and deoxygenated blood occurs at 3 anatomic sites, i.e., patent

ductus arteriosus, atrial septal defect, and ventricular septal defect. One or all of these lesions can be present, and the degree of arterial hypoxemia depends on the degree of anatomic mixing.

The majority of neonates with TGA manifest marked cyanosis within the first day of life. Metabolic acidosis also evolves secondary to poor tissue oxygenation. A bidirectional shunt is always present in TGA to prevent total volume depletion of the circulating volume. A major determining factor of systemic arterial oxygen saturation is the amount of blood exchange between the circulations.

Left to right shunt is from pulmonary to systemic circulation.

Right to left shunt is from systemic to pulmonary circulation.

Systemic arterial oxygen saturation is more influenced by left to right shunt. Infants with complete TGA are susceptible to the development of pulmonary vascular obstructive disease. Systemic arterial hypoxemia, increased pulmonary flows, and pulmonary hypertension, all contribute to the development of pulmonary vasoconstriction.

12.3 Perioperative Management

PGE_1 Infusion: Maintain ductal patency with continuous intravenous infusion to increase pulmonary blood flow, increase left atrial pressure, and promote left-to-right mixing at the atrial level.

Note:

a) PGE_1 infusion is critical in patients with severe left ventricular outflow tract stenosis or atresia.

b) Prostaglandin therapy, however, may or may not benefit the patient with simple transposition of the great arteries (TGA) with an intact ventricular septum.

Balloon atrial septostomy: Depending on the degree of restriction at the atrial septum and the timing of operative repair, it is indicated in severely hypoxemic patients with inadequate atrial level communication and insufficient mixing.

Metabolic acidosis: It should be corrected with fluid replacement and bicarbonate administration.

Mechanical ventilation: May be necessary if pulmonary edema develops with severe hypoxemia.

Surgical procedure: The patient may require corrective surgery or palliation early in life.

12.4 Operative Procedures

The procedures vary with the age of the patient at presentation, associated cardiac anomalies, and technical expertise of a surgeon. Most full-term neonates with uncomplicated transposition of the great arteries can undergo the arterial switch procedure in one operation, with a minimal mortality.

12.4.1 TGA with IVS (Intact Ventricular Septum)

The arterial switch procedure is an ideal operation. It should be performed when the infant is younger than 4 weeks, as the left ventricle may not be able to handle the systemic pressure postoperatively, if it is left too long in the low-pressure and low-resistance pulmonary circuit.

If the infant is older than 4 weeks, the left ventricle may be prepared to handle the systemic pressure by banding the pulmonary artery and placing an aorto-pulmonary shunt.

If coronary artery anatomy (e.g., intramural coronary artery) is not suitable for arterial switch, and coronary artery translocation may not be feasible, the atrial level switch (Senning or Mustard procedure) is favored.

12.4.2 TGA with VSD

The arterial switch procedure with VSD closure is the preferred operation. If coronary artery anatomy is such that arterial switch operation is inadvisable, and the VSD is large and nonrestrictive, Rastelli-type intracardiac repair may be feasible (see below).

If the infant has excessive congestive heart failure (with growth failure), it may be advisable to either follow with reparative surgery, or if it is not feasible, band the main pulmonary artery and place an aorto-pulmonary shunt during the newborn period to restrict pulmonary blood flow.

12.4.3 TGA with VSD and LVOT Obstruction (Pulmonary Stenosis or Atresia)

The arterial switch operation may not be feasible due to LVOT obstruction. If the VSD is nonrestrictive and not too remote from the aorta, the Rastelli intracardiac repair can be done. As Rastelli procedure necessitates a conduit from the right ventricle to the pulmonary artery, delaying the repair until the infant is older and larger may be preferable. In these patients, an aorto-pulmonary shunt may be done during the newborn period to establish adequate pulmonary blood flow while waiting.

12.4.4 TGA with VSD and Pulmonary Vascular Obstructive Disease

These patients are not appropriate surgical candidates because of progressive increase in pulmonary vascular resistance.

12.5 Postoperative Management

The postoperative course following arterial the switch surgery can be variable. The hospital stay averages one to two weeks.

12.5.1 Hemodynamic Management

Myocardial dysfunction is, at times, encountered and may be related to myocardial protection strategy and / or technical flaws in the insertion of coronary artery buttons in the neo-aorta.

12.5.2 Left Ventricular Dysfunction

It occurs if operative procedure is performed late, following regression of neonatal pulmonary vascular resistance.

12.5.3 Systemic Right Ventricular Dysfunction

It may occur after Mustard or Senning or for the congenitally corrected transposition and manifests in the later postoperative period. Intracardiac pressures following surgery should be normal. Arterial oxygen saturation should be normal.

12.5.4 Invasive Pressure Monitors

Arterial, central venous catheter, and LA (left atrial) catheters.

An oximetric catheter may be placed in PA pulmonary artery) to monitor cardiac output.

12.5.5 Vasoactive Drug Infusions

Dopamine or dobutamine, epinephrine, milrinone, nitroprusside, phenoxybenzamine, and nitroglycerin (see Section I Chapters 4 & 16).

12.5.6 Postoperative Bleeding

Lengthy great vessel suture lines may predispose to postoperative bleeding.

12.5.7 Cardiac Arrhythmias

Cardiac arrhythmias: Rarely are encountered.