Chapter

65

Transposition of the Great Arteries

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A 26-year-old has a full-term delivery of a 3.8 kg male baby that has an in-utero fetal diagnosis of transposition of the great arteries (TGA). Apgar scores are 4 and 6 (at 1 and 5 min). You are called upon for help.

What Is Transposition of the Great Arteries (TGA)?

TGA is a congenital cardiac malformation accounting for 5–7% of congenital cardiac anomalies. The basic abnormality in TGA is that the aorta arises from a morphologic right ventricle, and the pulmonary artery arises from a morphologic left ventricle (Figure 65.1). The common clinical entity of "simple" TGA is associated with situs solitus atria, concordant atrioventricular, and discordant ventriculo-arterial alignment. The anatomic derangement results in systemic venous blood flow predominantly to the aorta and pulmonary venous blood to the pulmonary artery creating a circulation in parallel. The deoxygenated and oxygenated blood need to mix at atrial, ventricular, or ductal level to be compatible with life. This physiologically uncorrected entity of TGA is fatal without treatment with mortality of 30% in the first week of life and 90% mortality within the first year.

What Are Oxygen Saturations in TGA?

Oxygen saturations greater than 85% on room air are reassuring; however, if the baby is severely cyanotic it is concerning as the baby may have pulmonary hypertension or poor mixing of the oxygenated and deoxygenated blood secondary to a restricted atrial septum.

What Is the Difference between D- and L-TGA?

In D-TGA the aorta is anterior and rightward, or dextroposed, and the circulations are parallel. In L-TGA, the circulation is in series and is often referred to as "corrected transposition" or "ventricular inversion." Anatomically, there is atrio-ventricular and ventriculo-arterial discordance which causes the deoxygenated venous return coming to the right atrium to get to the pulmonary artery but via a morphological left ventricle. The oxygenated pulmonary venous blood returns to the aorta via the morphological right ventricle.

What Is the Status of the Atrial Septum?

The atrial septum is very important in patients with D-TGA. This is the site of mixing of blood and allows the patient to have a systemic circulation with improved oxygenation. Secondly, the nonrestrictive atrial septum allows the decompression of the left atrium and a decrease in left atrial pressures.

What Is the Role of the PDA in This Congenital Anomaly?

The size and patency of the PDA is important (in the setting of a restricted atrial septum) in determining the degree of mixing of systemic and pulmonary circulations, and thereby survival. The patency of the PDA becomes less important if the atrial septum is non-restrictive. Hence it is not unusual to discontinue the prostaglandin after an atrial septostomy.

What Other Anomalies Occur with TGA?

No other coexisting defects may be present in nearly half of the patients with TGA. A persistent PDA or patent foramen ovale (PFO) may be the only anomaly. However, in about 40–45% of patients with TGA, a VSD is the most common anomaly. A combination VSD with significant left ventricular outflow tract obstruction (LVOTO) is seen in about 10% of patients. The presence of these coexisting defects along with valvular anomalies may alter surgical planning.

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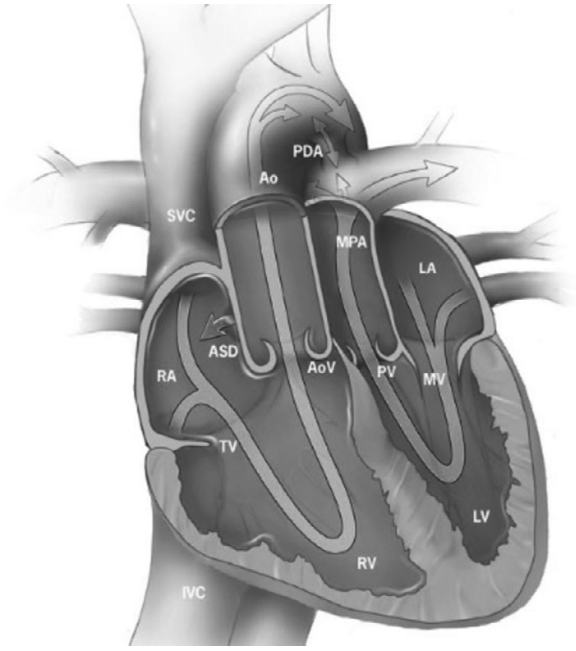

Figure 65.1 Illustration of transposition of the great vessels. RA and LA, right and left atrium; RV and LV, right and left ventricles; SVC and IVC, superior and inferior vena cava respectively; TV, MV, PV, AoV, tricuspid, mitral, pulmonary, and aortic valves respectively; MPA, main pulmonary artery; PDA, patent ductus arteriosus; ASD; atrial septal defect. Image courtesy of the Centers for Disease Control and Prevention, USA

The VSD may be small or large and located anywhere in the septum. Conoventricular or perimembranous VSDs account for 33% followed by malalignment for 30% and muscular defects accounting for 27%.

Is Pulmonary Stenosis Seen in Association with TGA?

Usually TGA with intact septum does not have pulmonary stenosis (PS).

Patients with TGA/VSD may have PS and can be extremely cyanotic.

How Is the Diagnosis of TGA Confirmed?

Echocardiography is the mainstay in confirming the diagnosis of d-TGA. The presence of a PDA, interatrial septal communication or VSD and other anomalies should be confirmed. These findings are important in surgical planning.

What Is the Difference in Circulation Physiology between a Normal Neonate and a Neonate Born with TGA?

The anatomic derangement results in deoxygenated systemic venous blood flow predominantly to the aorta and oxygenated pulmonary venous blood to the pulmonary artery creating a circulation in parallel. This means that the deoxygenated blood returns via the aorta back to the body and the oxygenated blood returns to the pulmonary circulation for re-oxygenation, causing a complete physiological right-to-left (to systemic circulation) and left-to-right shunt (to pulmonary circulation). The deoxygenated and oxygenated blood need to mix at the atrial, ventricular, or ductal level to be compatible with life. This mixing allows some deoxygenated blood to get oxygenated and some oxygenated blood to return to the systemic circulation.

How Is Survival Possible in a Parallel Circulation as Noted in TGA?

A parallel circulation is incompatible with life. There has to be mixing at the atrial, ventricular, or ductal level for the patient to survive. This physiologically uncorrected entity of TGA is fatal without treatment with mortality of 30% in the first week of life and 90% mortality within the first year.

What Are the Immediate Measures in a Neonate Born with a Confirmed Prenatal Diagnosis of TGA?

Room air oxygen saturations are one of the most important determining factors for management of the neonate. Umbilical lines may be placed as part of initial management. If the neonate is cyanotic, an atrial septostomy can be performed in the cardiac catheterization laboratory or at the bedside using echocardiography.

What Is a Typical ABG in d-TGA?

A typical arterial blood gas in room air reveals systemic pO_2 values <30 mm Hg that remain below 35–40 mm Hg during 100% oxygen administration, suggesting intracardiac mixing as the etiology of cyanosis versus a pulmonary etiology. In addition, in d-TGA the pCO_2 is also low as there is a high pulmonary blood flow which washes out the CO_2. The pH is also usually normal. In d-TGA there is no

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paucity of pulmonary blood flow, but it is oxygenated blood that keeps returning back to the pulmonary circulation. On the contrary, the systemic circulation receives deoxygenated blood and thus metabolic acidosis ensues due to anaerobic metabolism.

What Data on Echocardiogram Would Help Decide Medical Versus Interventional Management?

The status of the interatrial septum is the most important parameter in deciding further course of management.

What Is the Medical Management in the Newborn with TGA?

Treatment with prostaglandin to keep the ductus patent is the mainstay of medical management. Correction of metabolic acidosis and inotropic support is sometimes necessary with the goal of maintaining oxygen saturations between 75% and 85%.

What Are the Complications Associated with the Administration of Prostaglandins?

Prostaglandins can cause apnea, fever, seizures, and skin blanching.

What Is the Role of Mechanical Ventilation in This Medical Management?

Ventilation may need to be supported if the apneic spells are frequent and prolonged. The patient should be mechanically ventilated if a septostomy is performed.

What Is the Purpose of the Balloon Atrial Septostomy (BAS)? How Can It Be Performed?

The communication between the left and right atrium created by the atrial septostomy will allow for mixing of oxygenated and de-oxygenated blood.

The procedure can be performed at bedside or in the catheterization laboratory. In babies less than 48 h old, the umbilical vein can be used successfully for BAS. Percutaneous needle and catheter sheath placement via femoral vein access are other alternatives.

The catheter is advanced into the left atrium or pulmonary vein, across the foramen ovale. The catheter tip location should be confirmed – most readily by echocardiography (at-bedside–performed procedures) or by fluoroscopy (in the catheterization laboratory). The balloon is inflated with dilute angiographic contrast medium to approximately 15 mm in diameter, and then rapidly withdrawn across the septum with a short sharp tug. The procedure is repeated several times with increasing balloon volumes until balloon inflation at the level of the septum meets with minimal resistance. On echocardiography an interatrial defect of 5–6 mm represents an adequate BAS and palliation.

Complications such as tears of the atrial wall, pulmonary veins, and inferior vena cava (IVC) are uncommon. Temporary heart block which is self-limiting rarely happens and may rarely need long-term pacing.

BAS improves neonatal survival to more than 95%. However, centers that perform early arterial switch choose to perform BAS only in cases with profound hypoxemia or when surgery is delayed for other reasons.

What Are the Surgical Options for the Neonate with TGA?

If the baby is a few days old and has no other extra cardiac anomalies, the arterial switch operation, or Jatene procedure, is the surgical procedure of choice.

If the patient presents late or with other reasons to avoid cardiopulmonary bypass (CPB), the patient can be managed for a few months medically with an atrial septostomy. The patient would then be a candidate for an "atrial switch procedure" like Mustard or Senning where an atrial baffle is created to redirect the deoxygenated blood to the left ventricle, which is connected to the pulmonary artery. The atrial switch operation leaves the morphological right ventricle as the systemic ventricle.

What Are the Names of the Surgical Procedures and How Do They Differ from Each Other?

Arterial switch: Jatene's procedure (1975) – creates left ventricle as the systemic chamber and anatomically provides corrective repair.

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Atrial switch: Senning (1959) or Mustard (1964), which baffles the venous return to the left ventricle and serves as the pulmonary ventricle, leaving the right ventricle as the systemic ventricle. This results in physiologic correction however with the left ventricle functioning as the pulmonary ventricle and the right ventricle as the systemic ventricle.

In the Senning procedure the atrial baffle is created from the right atrial wall and atrial septal tissue. No extrinsic tissue or materials are used.

The Mustard operation on the other hand involves resection of the atrial septum and creation of a baffle from synthetic material.

Both these procedures are performed usually between 1 month and 1 year of age. Surgical mortality from these procedures is usually between 1% and 10%.

Rastelli operation (1969): This surgical option is usually reserved for TGA with pulmonary outflow tract obstruction and VSD. The procedure entails using the VSD as part of the LVOT and placement of a baffle within the RV directing oxygenated blood from the VSD to the aorta. The pulmonary valve is oversewn and a conduit is inserted between the RV and PA. A subaortic and large VSD is suitable for this procedure as surgical enlargement carries a large risk of causing complete heart block. The biggest advantage with this procedure is that the LV becomes the systemic ventricle, however an important limitation is that multiple reoperations are needed during the patient's life.

What Prompts Correction versus Palliative Procedure?

Age. If the patient is older than six to eight weeks, then the left ventricle is de-conditioned as it is no longer pumping to the systemic circulation but instead to the pulmonary circulation. The pulmonary circulation is at a lower pressure than the systemic circulation so performing an arterial switch at a later age does not allow the LV to generate systemic pressure. The patient may need to undergo pulmonary artery banding to prepare the LV prior to a switch.

The presence of other extra-cardiac anomalies may preclude an arterial switch operation.

Other conditions like RSV infection and MRSA pneumonia may preclude CPB.

What Are Some of the Anesthetic Considerations Preoperatively in This Neonate Scheduled for TGA?

These patients are usually monitored and managed in the ICU setting. It is important to perform a complete assessment of the child for any extra cardiac anomalies which can alter further management. A review of the arterial blood gases since birth should be done to identify current ventilation, oxygenation, and acid base status. A neutral acid base status with saturations in the mid 80% is preferred for a balanced circulation hence avoiding systemic acidosis or pulmonary overcirculation.

These patients may be supported on mechanical ventilation secondary to increasing apneic spells by the institution of prostaglandin infusions to keep the PDA open for systemic circulation. An assessment of the umbilical lines if present should identify the correct depth of catheter insertion and presence of free aspiration of blood.

What Are Some of the Risk Factors for an Increased Perioperative Mortality?

A restrictive PFO or ASD, persistent pulmonary hypertension, prematurity, low birth weight and a delayed diagnosis all tend to increase mortality.

What Is the Strategy for Monitoring in These Patients during Surgical Correction of TGA?

The presence of a fresh umbilical stump makes it conducive for placement of invasive monitoring lines both venous and arterial. Normally the umbilical cord is composed of a gelatinous material – Whartons jelly – two umbilical arteries, and one umbilical vein. These blood vessels can be accessed under sterile conditions for central venous access and invasive arterial pressure monitoring. Central venous access under ultrasound guidance via the internal jugular or femoral route is another alternative.

What Are Some of the Important Intraoperative Anesthetic Considerations?

Almost all of these infants will present to the OR with an intravenous access – central (umbilical) or

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peripheral. A narcotic-based induction technique followed by a balanced anesthetic with narcotics and inhalational agents is often a prudent choice with better control of hemodynamics as well as blunting of the stress response.

Occasionally profound arterial desaturations may be seen during induction of anesthesia and positive pressure ventilation in the patient with TGA/IVS. It is often related to the sudden suppression of endogenous catecholamines, along with the decreased preload due to positive pressure ventilation and decreased myocardial contractility due to the high dose narcotics. Inadequate ventilation may cause hypercarbia and pulmonary hypertension.

Ensuring adequate ventilation, oxygenation, anesthetic depth and volume status are the steps to ensuring recovery from these critical moments failing which cardiopulmonary bypass may need to institute rapidly.

Is the Cardiopulmonary Bypass Different for Correction of TGA?

The conduct of CPB is unique to each institution and may include flow rates of 200 mL/kg/min, moderate hypothermia of 28–32°C, blood cardioplegia, and modified ultrafiltration at the termination of CPB. The use of antifibrinolytics (aminocaproic acid) is useful in control of coagulopathies.

What Are Some of the Critical Issues Soon after Termination of Cardiopulmonary Bypass?

As soon as CPB is terminated, global ventricular function and regional myocardial wall motion abnormalities should be assessed by transesophageal echocardiograph (TEE). The presence of regional wall motion abnormalities (RWMAs) may alert to kinking or spasm of the coronary ostia which may require repair of anastomosis and/or infusion of nitroglycerin to maximally dilate the coronaries.

The presence and persistence of arrhythmias is an indicator of abnormal myocardial perfusion and coronary artery problems must be ruled out.

Neonates with TGA/intact ventricular septum (IVS) usually will not tolerate acute and significant changes in preload and afterload due to a deconditioning of the LV. Transfusion of blood and vasodilators should be carefully titrated to intracardiac filling pressures (LA or RA) along with maintenance of systolic blood pressure in the range of 50–70 mmHg and RA pressures in the range of 5–6 mmHg.

Chest closure may sometimes become difficult due to myocardial edema, hemodynamic instability, coagulopathy, and need for transfusion of blood products.

Is Inotropic Support Mandatory for the Termination of Bypass?

Inotropic support is needed in situations where global ventricular function is depressed and coronary arterial abnormalities have been excluded. Infusions may include either or a combination of dopamine 3–10 mcg/kg/min, epinephrine 0.03–0.05 mcg/kg/min or milrinone 0.25–0.75 mcg.kg/min. Nitroglycerin infusion at 1–2 mcg/kg/min may be added to aid in coronary vasodilation and relief of increased afterload.

Increasing dosage of inotropic support with increasing systemic acidosis and hemodynamic instability warrant re-exploration and correction of the coronary anastomosis.

Is There a Need for Intraoperative Transesophageal Echocardiography?

Intraoperative TEE is useful in the assessment of ventricular function and regional wall motion abnormalities after switching of the great vessels and reimplantation of coronaries to the neoaorta. TEE is also useful in the assessment of de-airing of the ventricles after completion of the surgical repair and cessation of cardiopulmonary bypass. TEE is also useful in assessment of the anastomotic sites of the neoaorta and neopulmonary artery.

What Are the Immediate Postoperative Concerns in the Intensive Care Unit after Correction of TGA?

Arrhythmias, depressed ventricular function, and ongoing coagulopathy are critical issues during the early postoperative period. Cardiac catheterization for assessment of coronaries and surgical reexploration is warranted if myocardial function continues to be depressed or deteriorating. Circulatory support with extracorporeal circulation should be warranted if hemodynamic instability continues.

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What Are the Common Complications after an Arterial Switch Operation (ASO)?

Arrhythmias, LV dysfunction, sudden cardiac death, pulmonary artery stenosis, and neoaortic valve regurgitation are common late sequelae after ASO. Symptoms of myocardial ischemia are atypical due to the denervation of the heart at the time of the ASO. It is therefore important to be aware of this complication and remain vigilant.

Suggested Reading

Boris JR. Primary care management of patients with common arterial trunk and transposition of the great arteries. *Cardiol Young*. 2012;22 (6):761–7. PMID: 23331600.

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