Perioperative anesthetic management of transposition of great arteries: a review

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Transposition of great arteries (TGA) comprises 5–7% of all CHDs. It is characterized by atrioventricular concordance and ventriculoarterial discordance, resulting in the systemic and pulmonary circulations as parallel instead of the normal in-series circulation. Survival of the baby depends on mixing of blood between these two circulations either with an atrial septal defect, ventricular septal defect, or at the great arterial level via patent ductus arteriosus. Therefore, the clinical manifestation is highly variable and influenced by the presence or absence of these associated anomalies. Patients with TGA without mixing of blood present with cyanosis and acidosis and are hemodynamically compromised soon after birth and require resuscitation to re-establish connection between parallel circuits by reopening the ductus with intravenous prostaglandin (0.05–0.1 μg/kg/min) or establishing interatrial flow with balloon atrial septostomy. In addition, patients may require inotropic support, ventilator support, or extracorporeal membrane oxygenation in extreme cases with refractory cardiorespiratory decompensation for survival or as a bridge to definitive therapy. TGA is uniformly fatal in the infant period, with 30% mortality in the first week of life, and 50% within the first month, and 90% in the first year of life if untreated. Fortunately, modern medical and surgical management techniques have resulted in 90% of patients living into adulthood, typically with a vigorous quality of life. Currently, the definitive corrective surgery is the arterial switch operation (ASO), as a single-stage procedure with excellent short-term and long-term outcomes. The overall perioperative survival following ASO is more than 90%. Long-term and arrhythmia-free survival is ~97% at 25 years. All standard general anesthetics can be used safely for perioperative management, and mortality owing to anesthetic management has not been witnessed. This systematic review describes the definition and etiology of TGA, clinical presentation, pathophysiology, brief current surgical approaches, anesthetic and cardiopulmonary bypass management, and postoperative course of a patient with TGA undergoing ASO.

Keywords:
TGA, arterial switch operation, atrial switch operation, ECMO, CPB, rastelli procedure, Nikaidoh procedure, prenatal diagnosis

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Introduction

Transposition of great arteries (TGA) is one of the most common cyanotic congenital heart defects diagnosed at birth. It is ventriculoarterial discordance, in which the aorta arises from the morphologic right ventricle and the pulmonary artery arises from the morphologic left ventricle (LV). Anatomically, it is classified as dextro (D-TGA) and levo transpositions (L-TGA), based on the relationship of the pulmonary artery with the aorta in the anomalous heart. TGA occurs in 5–7% of all congenital heart defects and almost 20% of all cyanotic cardiac diseases. D-TGA is the second most common congenital cardiac defect noted at birth, affecting 1 : 3500–5000 live births with a male : female ratio of 3.2 : 1.3 [1-8]. Consequently, D-TGA results in the lethal hemodynamic pattern of two independent and parallel running circulatory circuits, instead of in the series [9]. Complete parallel circuits are incompatible with life and mandate a patent ductus arteriosus (PDA) or atrial septal defect (ASD) or ventriculoseptal defect (VSD) for mixing of oxygen-rich and oxygen-poor blood for survival [5]. Adjuvant therapies, specifically the Rashkind balloon atrial septostomy (BAS), prostaglandin infusion, mechanical ventilation, and inotropic agents, have the potential to improve systemic arterial oxygen saturation and tissue oxygen delivery. However, adjuvant therapy is short term until

a definitive treatment in the form of arterial switch operation (ASO) can be performed. TGA can result in acute cardiorespiratory decompensation and death within the first 48 h of life if the diagnosis is missed. D-TGA has a 90% mortality rate in the first year of life if untreated; however, more than 95% survive for 5–25 years after surgery [5]. Recent evolutions in preoperative evaluation, patient selection, preoperative preparation, medical and surgical advances, perfusion techniques, and postoperative care have spectacularly changed the outcome of this lesions. Currently, the ASO is the mainstay of management of these patients with excellent results [10]. Surgical intervention requires the cardiac anesthesiologist to fully understand the pathophysiology, suitability for ASO, aims of treatment, surgical procedures, and postoperative consequences to form the perioperative management plan. Perioperative anesthetic considerations include comprehensive preoperative cardiac evaluation for suitability of ASO and management of hypoxemia and ventricular dysfunction. Alterations in pulmonary vascular resistance (PVR) and systemic vascular resistance (SVR) are to be prevented especially at the time of induction and off cardiopulmonary bypass (CPB), and cardiac output should be maintained using balanced opioid-based anesthetic techniques.

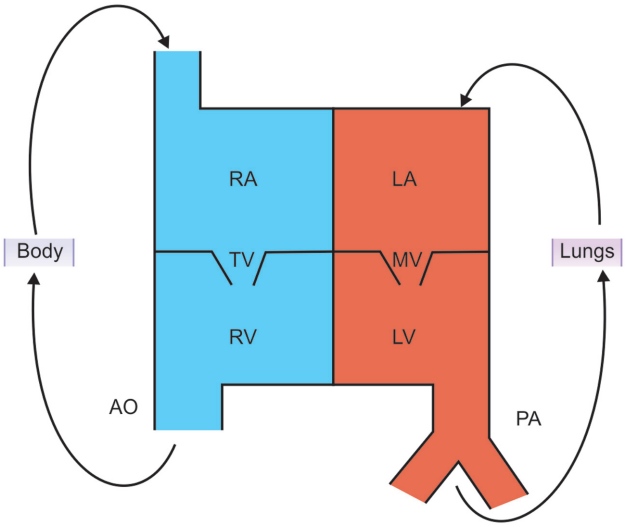
Methods

Electronic searches for this systematic review included PubMed, Medline, research gate, Google, and the Cochrane database up to March 2022. Selection criteria were case reports, case series randomized studies, meta-analysis, reviews, and clinical guidelines of TGA surgery mainly ASO repair. The primary focus was on clinical presentation, pathophysiology, initial management, surgical decision making, anesthetic management for ASO, various surgical options, postoperative management, and early and long-term outcomes of ASO.

Embryology, definition, differential diagnosis, and etiology of transposition of great arteries

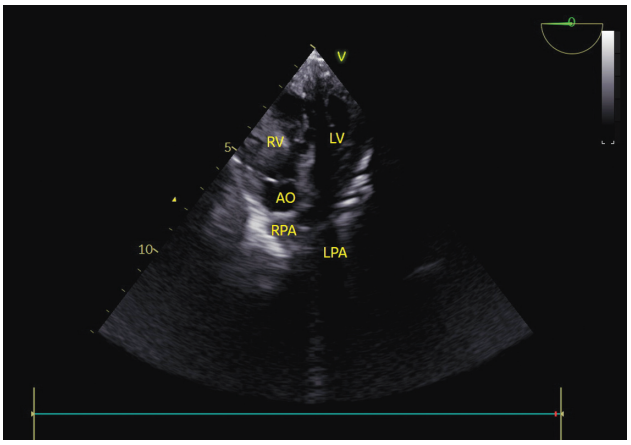
TGA develops from an embryological discordance between the aorta and pulmonary trunk. During heart development, the conotruncal septum spirals toward the aortic sac thus dividing the truncus arteriosus into the pulmonary and aortic channels. These channels then become the pulmonary arteries and aorta, respectively. TGA occurs when the conotruncal septum fails to follow its spiral course and instead forms in a linear orientation. Consequently, the aorta arises from the right ventricle and the pulmonary trunk arises from the LV. TGA is divided into dextro-looped (D-TGA) and levo-looped (L-TGA), also known as congenitally corrected TGA (CCTGA). It is based on whether the atria and ventricles are concordant (D-TGA) or discordant (L-TGA) [1,8]. In D-TGA, the right atrium (RA) is connected to the right ventricle (RV), which is further connected to the aorta, whereas Left atrial (LA) is connected to the LV which is further connected to the PA, and RV is placed to the right of the LV, and the aorta is anterior and to the right of the PA (Figs 1 and 2). This relationship of the ventricles to their respective great arteries is termed as ventriculoarterial discordance, resulting in a parallel circulation. However, CCTGA is a rare anomaly, with an incidence of 0.03 per 1000 live births, accounting for ~0.05% of all forms of congenital heart diseases (CHD), in which the morphological RA is connected to a morphological LV across the mitral valve (atrioventricular discordance) and the LV then is connected to the PA (ventriculoarterial discordance). The morphological LA is connected to the morphological RV across the tricuspid valve (TV), and the morphological RV is then connected to the aorta. Deoxygenated systemic blood enters the right atrium, goes through the morphological LV, and enters the pulmonary circulation via the PA. Oxygenated blood from the pulmonary circulation then enters the LA, goes through the morphological RV, and

Figure 1



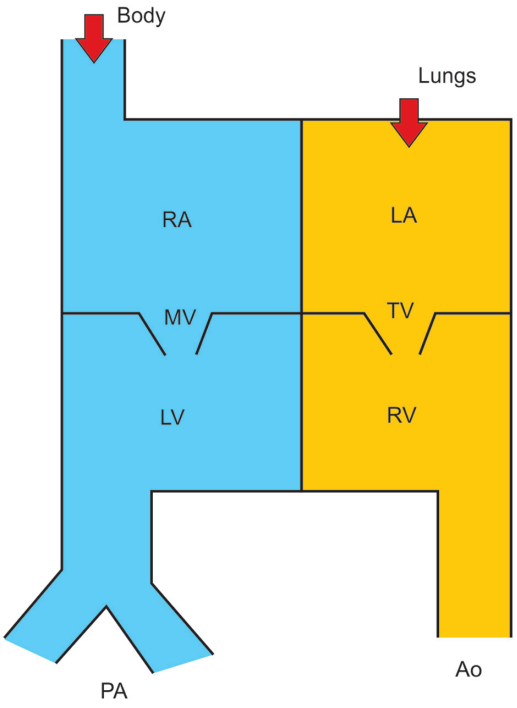
RA is connected to the RV and aorta is attached to the RV. The LA is connected to the LV via MV and PA originates from the LV. The deoxygenated blood from the body comes to RA via SVC and IVC and goes to RV through TV and again recirculates to the body by aorta. The oxygenated blood from lungs goes to LA, from LA to LV via MV, and recirculates to lungs via PA. Therefore, the deoxygenated blood goes back to body and the oxygenated blood goes back to the lungs.

Figure 2



Deep TG LAX. Two-dimensional TEE image displays that PA originates from LV and divides into RPA and LPA with acute angles, and AO is connected to the RV (Labeled) suggestive of ventriculoarterial discordance of TGA. AO, aorta; LPA, left pulmonary artery; LV, left ventricle; RPA, right pulmonary artery; RV, right ventricle; TGA, transposition of great arteries.

Figure 3



Diagrammatic presentation of CCTGA or L-TGA with double discordance, that is, ventriculoarterial and atrioventricular. The figure demonstrates that oxygenated blood reaches to RV and pumped to the body by the aorta and deoxygenated blood reaches to the LV and further to the lungs by the PA for oxygenation (labeled). AO, aorta; IVC, inferior vena cava; LV, left ventricle; MV, mitral valve; PA, pulmonary artery; RA, right atrium; RV, right ventricle; SVC, superior vena cava; TGA, transposition of great arteries; TV, tricuspid valve;.

enters systemic circulation via the aorta (Fig. 3). Hence, the CCTGA is described as 'double discordance' resulting in normal physiology [11-13]. TGA is traditionally categorized into three subtypes: TGA with intact ventricular septum (TGA-IVS) (70%), TGA with ventricular septal defect (TGA-VSD) (25%), and TGA with VSD and left ventricular outflow tract obstruction (TGA-VSD-LVOTO) (5–10%) [14].

Differential diagnosis of TGA includes tetralogy of Fallot, tricuspid atresia, truncus arteriosus, and total anomalous pulmonary venous connection. There are some other lesions which need to be differentiated from TGA like single ventricle, hypoplastic left heart syndrome, Taussig–Bing anomaly (DORV plus TGA), single atrium, and pulmonary stenosis or atresia [15]. The etiology of TGA is unknown, but factors like viral infection or German measles to the mother during pregnancy, poor nutrition, alcoholic drinking during pregnancy, poorly controlled diabetes mellitus, and old mother (>40 years) may increase the risk of this condition. There is also a strong association with family history of TGA and Down's syndrome in the baby [1,7].

Pathophysiology of transposition of great arteries

In a baby with a normal heart after birth, the right side of the heart pumps deoxygenated blood to the lungs through the PA and the left side of the heart pumps oxygen-enriched blood to the body through the aorta. In TGA, the oxygenated blood recirculates within the pulmonary circuit via the LV and pulmonary trunk, whereas deoxygenated systemic blood recirculates to the body via the right ventricle and aorta (Fig. 1). These two parallel circuits without any mixing of blood result in systemic cyanosis, acidosis, and rapid decompensation, leading even to death. The degree of cyanosis in the first-day baby depends on the amount of mixing of blood via an ASD or VSD or PDA or big major aorto-pulmonary collateral artery, and adequate mixing is an important determinant of the clinical presentation, preoperative medical management, and surgical timing. Therefore, TGA without surgical or pharmacological interventions is associated with very high mortality, within the first year of life [4]. Unoperated TGA associated with ASD and/or VSD has an average life expectancy of 9 or 22 months, respectively [4]. Therefore, the diagnosis of TGA is an indication for treatment owing to scarce survival beyond the neonatal period if not corrected [16].

Neonates with TGA and adequate mixing of blood may be only mildly cyanotic at birth. Initially at birth, PVR is greater, or equivalent to the SVR, allowing partial mixing of blood between the parallel systemic and pulmonary circuits via the ASD or VSD or PDA. As PVR decreases by 2–6 weeks, the balance is

disturbed, thereby allowing increased shunting from the systemic to the pulmonary circuit, resulting in pulmonary congestion and developing symptoms of congestive heart failure, namely tachypnea, tachycardia, mild systemic cyanosis, lack of appetite, and growth retardation [17,18]. In contrast, most patients with an isolated CCTGA remain asymptomatic through infancy and childhood and even maintain normal biventricular function through early adulthood and can survive a normal life span [19]. However, patients with long-standing CCTGA may present with ventricular dysfunction, regurgitation of the tricuspid valve, arrhythmias, heart block, congestive heart failure, heart palpitations, dyspnea, fatigue, and fainting spells (syncope) [16].

Diagnosis of transposition of great arteries

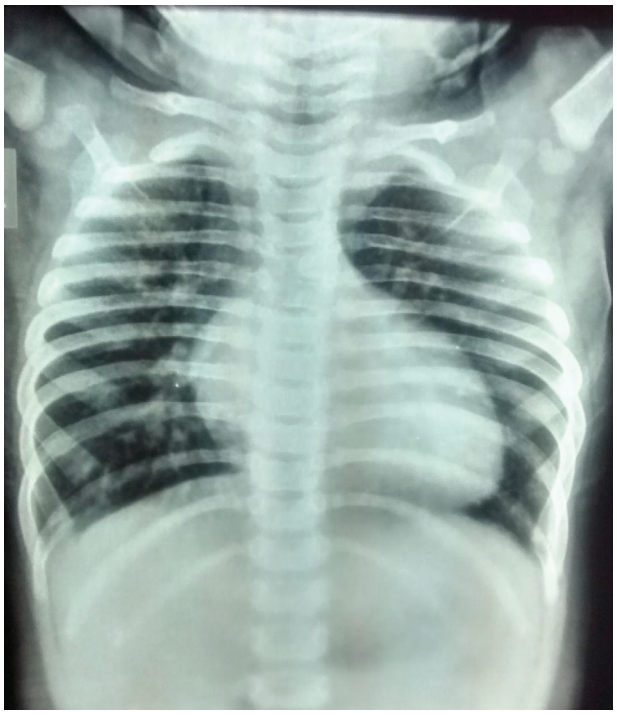
Prenatal diagnosis

The TGA can be diagnosed accurately before birth if the fetal heart is screened at the time of the obstetric anomaly scan (18–22 weeks of gestation). In addition, the type of repair likely to be required after birth can also be well predicted beforehand. It has been observed that the four-chamber view alone is insufficient to diagnose the TGA-IVS, but inclusion of the outflow-tract views at the time of the obstetric fetal anomaly scan results in significant improvement in prenatal detection of the TGA, that is, up to 50% [20-22]. Therefore, it has been recommended that in addition to the four-chamber view, the cardiac outflow tract views should also be included as part of the obstetric screening scan to determine the TGA [23]. If TGA is detected and diagnosis is confirmed, the parents should be informed and counseling should also be provided by a fetal cardiology specialist and other related health professionals (fetal medicine specialists, obstetricians, pediatric cardiac surgeons, and neonatologists) about the postnatal outcome [24].

Diagnostic features

Clinically, infants with TGA usually present with cyanosis immediately after birth. Cyanosis is more pronounced in the absence of adequate intercirculatory mixing. Occasionally, D-TGA with large VSD develops signs of congestive heart failure (tachypnea, tachycardia, diaphoresis, and loss of weight gain) over the first 3–6 weeks as pulmonary blood flow increases. ECG is typically normal. Chest radiograph may suggest a narrow superior mediastinum with small thymic shadow and anterior–posterior position of great vessels and an egg-shaped heart owing to enlarged RA and RV (Fig. 4). The main diagnostic modality is echocardiography, which establishes the diagnosis and identifies the presence of other associated anatomical defects and provides all the anatomical and functional information needed for the management. It can identify the aorta arising from RV and PA from LV and their interrelationship, that is, superimposed or side by side, which has significance during ASO. In addition, site, size, and direction of shunt can also be delineated, and the risk factors like anomalous course of coronary arteries, underdeveloped LV and LVOTO, and multiple VSDs can be identified. Pulmonary artery pressure, RV functions, mitral regurgitation, and LV size, thickness, mass, and function can also be assessed to decide the surgical approach and determine the surgical outcome. Echocardiography and coronary computed tomography angiography allow for a complete visualization of the origin and course of the coronary arteries. The common anomalies include single coronary artery (6%), circumflex arising from right coronary artery (RCA) (20%), left anterior descending (LAD) originating from RCA, and inverted or intramural course of coronaries. These coronary artery patterns can affect the surgical repair of D-TGA and require the technical modification in coronary artery reimplantation [25]. Cardiac catheterization is utilized when echocardiography does not provide enough information to make a diagnosis. It confirms the amount of blood mixing, coronary artery anatomy, and pulmonary vascular resistant and is also used as

Figure 4



Chest radiograph suggests a narrow superior mediastinum with small thymic shadow and anterior–posterior position of great vessels, and egg-shaped heart owing to enlarged RA and RV.

a part of urgent palliation such as BAS and ductus stenting.

Initial management of D-transposition of great arteries

The assessment should include a detailed review of the patient's course in the hospital. The initial management of newborns with TGA should focus on stabilization, optimization of mixing of systemic and pulmonary circulations, arterial oxygen saturation, oxygen delivery, maintenance of adequate systemic perfusion, and correction of acidosis [26]. Neonates with profound hypoxemia (partial pressure of arterial oxygen <25 mmHg and SaO_2<60%) require urgent attention. Most hypoxic newborns are started on PGE1 (0.05–0.1 μg/kg/min) to maintain arterial duct patency and therefore increase systemic oxygen saturation. Patients on PGE therapy have a lower postoperative inotrope score [27]. However, prostaglandin therapy may cause apnea spells, seizures, systemic hypotension, inhibition of platelet aggregation, peripheral edema, and unexplained fever, which may require intubation and volume expansion with normal saline. A subset of patients with poor intracardiac mixing (restrictive PFO or IVS) experience markedly increased pulmonary blood flow with PGE alone, leading to LA hypertension, pulmonary overcirculation, and systemic hypoperfusion. These newborns, plus those with both hypoxia and acidemia, require urgent BAS (4–5 mm), surgical septostomy, or blade atrial septostomy under echocardiography guidance in ICU to provide intracardiac mixing and balance of the circulations. The current practice is to perform BAS in neonates who have both echocardiographic evidence of a restrictive atrial septum and hypoxia or instability that is unresponsive to other interventions [28-30]. It dramatically improves the arterial saturation and hemodynamic and clinical status of the patient. However, BAS has also been reported to have complications like arrhythmias, vascular or atrial trauma, and cardiac tamponade, and seizures and embolic stroke [31]. Once adequate intercirculatory mixing is established with BAS, PGE, or both, the patients generally stabilize quite quickly, yet early surgical repair remains indicated to lessen the risk of neurologic injury from periods of marginal oxygenation and perfusion.

In addition, 1–12% neonates with TGA-IVS develop persistent pulmonary hypertension of the newborn (PPHN) before surgical repair [9,32]. In these infants, the early diagnosis of PPHN and treatment with iNO may improve outcome. In the presence of moderately large atrial communication and PPHN, treatment with iNO should be considered before BAS. iNO is started initially at 10 ppm and then increased to 20 ppm [33]. The iNO treatment should be gradually withdrawn and discontinued after 12 h, as abrupt withdrawing can result in severe rebound PAH and life-threatening hypoxemia. Therefore, iNO should be continued postoperatively via an endotracheal tube or face mask, and weaning should be done slowly with increasing oxygen supplementation along with consideration of the administration of oral or intravenous pulmonary vasodilators before discontinuation of iNO. Some patients with TGA-IVS with restrictive foramen ovale and/or closure of the ductus arteriosus present with extreme cyanosis, severe hypoxia, and acidosis early after the birth and remain unresponsive to ongoing therapy (PGE and BAS). Such neonates may present at the most severe end of the spectrum, that is, progressive cardiogenic shock with a high risk of end-organ injury, or even cardiac arrest. In this condition, patients are unsuitable to undergo ASO immediately and require extracorporeal membrane oxygenation (ECMO) support preoperatively [34]. Lorts and Krawczeski [35] have described that ECMO has been a successful bridge to corrective surgery with excellent outcomes. Once the infant is stabilized, corrective surgery is optimally performed in the first weeks of life.

Training and assessment of left ventricular preparedness for a rapid arterial switch operation

Once the PVR starts falling at about fourth week after birth, the LV mass in TGA-IVS starts reversible decaying. On the contrary, in patients with TGA and RVOT obstruction, the LV mass is maintained and allows an early ASO. In patients with unprepared LV, a PA banding combined with a modified Blalock–Taussig shunt using a 3–3.5 mm PTFE tube is used according to the weight of the patient (<3 kg or >3 kg), and then an ASO has been recommended after an interval depending on the patient's age [i.e. 1–2 weeks in young infants (rapid two-stage ASO)]. The categorical indications for LV training before ASO include a combination of the following criteria: indexed LV mass less than 35 g/m^2 (on echocardiography or MRI), age well above 3 weeks, ventricular septal profile, with a banana-like LV shape on two-dimensional echocardiograms, absence of a PDA or LVOT obstruction, and LV to RV pressure ratio less than 0.6 [33]. The proposed criteria for a safe second-stage ASO include LV-to-RV pressure ratio more than 0.85, LV end-diastolic volume more than 90% of normal, LV ejection fraction more than 0.5, posterior wall thickness more than

4 mm and a predictive wall stress less than 120×10^3 dynes/cm^2, not more than mild MR, LVEDP less than 12 mmHg, LVESP more than 80% of systemic pressure, and LV mass more than 50 g/m^2. The Devereux formula for calculation of LV mass is in wide clinical use. The formula is stated as follows: 0.8 $\{1.04[(LVEDD+IVSd+PWd]^3-LVEDD^3)]\}+0.6$ g, where LVEDD, IVSd, and PWd represent LV, interventricular septal, and posterior wall thickness in diastole, respectively [36,37]. Some authors have suggested the optimal interval between stages I and II as 10 days (range, 5 days to 6 weeks) [38]. The PA banding can be used to make unprepared LV suitable for future successful ASO. Some authors have suggested PA banding combined with induced patency of the ductus arteriosus, obtained by either PGE1 infusion or ductal stenting, depending on the anticipated duration of the interim period [38]. Alternatively, LV preparedness may be assessed by a 'provocative' pulmonary artery banding: if tolerated by the LV for up to 15–30 min, a primary ASO is undertaken [39].

Surgical options for repair

Arterial switch operation

Currently, the ASO with excellent operative survival is the gold standard treatment for the neonates with TGA and IVS. It achieves anatomical correction rather a physiological restoration of the circulation. In ASO, the aorta and PA are transected, the latter proximal to the bifurcation. The left and right coronary arteries are then resected together with a small patch known as coronary buttons. The coronary arteries are implanted to the neoaorta (formerly the main pulmonary artery). The transected great vessels are switched, creating neoaorta and neopulmonary trunk. The proximal transected pulmonary trunk is shifted posterior to the branch pulmonary arteries (The 'Lecompte maneuver') and anastomosed to the distal aorta, forming a neoaorta, and the proximal transected aorta is repaired with the pericardial patch at the site of coronary artery explantation. Subsequently, this is anastomosed to branch pulmonary arteries, forming a neopulmonary trunk. In addition, the ASD is also repaired using a pericardial patch [15,40]. On completion of the procedure, the LV receives oxygenated blood from the lungs, which it then pumps to the body. The RV pumps the systemic venous return to the lungs in the usual fashion. The definitive corrective procedure is performed between the neonatal period and the first 6 months of life. Some authors have suggested the third day of age as the ideal time for an ASO when LV mass is still adequate [41,42]. It has been reported that ASO can be safely performed within hours of the birth using autologous umbilical cord blood to prime the CPB circuit, as early ASO improves the outcome and reduces the costs [41].

Prêtre *et al*. [43], Fricke *et al*. [10], and Shim *et al*. [44] have reported that the coronary events related to myocardial ischemia have been the most common cause of early mortality.

Coronary artery-related myocardial ischemia often occurs owing to mobilization or anastomosis, twisting, kinking of the coronary arteries, and because of the myocardial edema. In addition, the multiple and extensive suture lines of ASO can lead to stenosis of aorta or pulmonary artery and excessive bleeding and tamponade. Myocardial dysfunction can also occur owing to long CPB and aortic clamp time. CPB time greater than 150 min is an independent predictor of both hospital and intermediate-term mortality of ASO [45]. CPB has well-known time-dependent deleterious effects, including a whole-body inflammatory response, metabolic changes, and fluid and electrolyte imbalance [46]. Global systolic and diastolic LV dysfunctions and low cardiac output occur often after 6 h of the ASO, and LV dilates and poorly tolerates volume. Following ASO, arrhythmias and cardiac dysfunction should raise suspicion of coronary insufficiency [47]. Hui *et al*. [48] and others have described even a baseline abnormal LV contractility in a significant proportion (61%, 19 of 31) of patients with TGA and that may persist in the postrepair period [41].

However, several authors have described that due to advances in surgical technique, diagnostic methods, anesthetic care, perfusion protocols, and preoperative and postoperative intensive care, the outcome for this lesion has improved significantly. Recent studies have demonstrated a hospital survival rate of more than 98% after ASO. Data from the United Kingdom's National Institute for Cardiovascular Outcomes show a nationwide 30-day mortality rate for the ASO at less than 3% with a 1-year survival rate of more than 96% [1,10]. Long-term and arrhythmia-free survival is excellent after effective ASO, although sequelae include chronotropic incompetence, RVOT obstruction, and neoaortic, pulmonary artery, and coronary artery complications [1,49]. Khairy *et al*. [1] have conducted a single-institution retrospective cohort study to assess cardiovascular outcomes after an ASO over 16 years. A total of 400 patients, including 154 (38.3%) with a VSD, 238 (59.5%) with an intact septum, and nine (2.3%) with a Taussig–Bing anomaly,

were followed for a median of 18.7 years. In perioperative survivors, overall and arrhythmia-free survival rates at 25 years were 96.7±1.8 and 96.6 ±0.1%, respectively, and 92.9±1.9% were free from adverse cardiovascular events. More recently, Raissadati and colleagues reported 25-year survival rates of 97.0% (95% confidence interval: 95.0–100.0%) among patients undergoing ASO [50,51].

Atrial switch operations

These were the palliative procedure of choice between 1960 and 1980s, known as Mustard or Senning repair. Intra-atrial baffle repair, as pioneered by Senning and Mustard, has radically altered the otherwise grim associated prognosis, allowing most patients with D-TGA to thrive well into adulthood [52]. In the Mustard procedure, a pericardial patch is used to baffle systemic venous flow to mitral valve and pulmonary venous flow to tricuspid valve. The Senning procedure has similar principles as the Mustard but uses the native atrial tissue to create a 'baffle' between the atria to redirect the deoxygenated vena caval blood through the mitral valve to the morphological LV and pulmonary arteries, and oxygenated pulmonary venous blood through the tricuspid valve into the morphological RV and aorta [53,54]. Both procedures result in a physiologic correction of the TGA, rather an anatomic correction, and leave the RV as a systemic ventricle. Although these procedures have excellent early survival (85–90%), but later on, they may be associated with the development of baffle stenosis obstructing the pulmonary veins or vena cave resulting in PAH, SVC, and IVC syndromes and low cardiac output. In long-term baffle leaks, atrial arrhythmias (atrial conduction defects, sick–sinus syndrome with brady and tachyarrhythmia, and atrial flutter), protein loosing enteropathy, tricuspid regurgitation, RV dilatation and ultimately RV failure, and sudden death are other postoperative complications [55-58]. Therefore, nowadays, Senning and Mustard procedures are only performed in those patients who have unswitchable anatomy owing to complex coronary abnormalities, late diagnosis, coexistent VSD with associated pulmonary hypertension, inadequate LV function, and unprepared LV [59]. Sometimes, the authors have witnessed a severe intratracheal bleeding after atrial switch repair leading to severe hypoxemia necessitating frequent gentle endotracheal suction and vigorous hand ventilation to expand the lungs and increase the functional residual capacity.

Rastelli procedure

D-TGA associated with VSD and some degree of LVOT obstruction is an indication for Rastelli procedure. In this procedure, the VSD is closed using a Gore-Tex patch in such a way that the oxygenated blood from the LV is directed to the aorta. The pulmonary valve is surgically closed and an artificial valve conduit is connected from the RV to the pulmonary bifurcations, allowing deoxygenated blood to travel to the lungs for reoxygenation [60]. The long-term complications include heart blocks and LVOT obstruction at the VSD patch as the child grows and others such as extra-cardiac conduit stenosis (with or without regurgitation), calcification, kinking, and aneurysm, and tunnel patch from the LV to the aortic valve may be complicated by leakage, obstruction, stenosis or aneurysm, and branch pulmonary artery stenosis biventricular dysfunction. Alsoufi *et al.* [60] have reported zero early mortality and mid-term results at 10 years suggested that 92% patients remain in New York Heart Association functional class I and class II. None of their patients had late arrhythmias or required heart transplantation.

Nikaidoh procedure

It is indicated in patients with TGA with VSD and pulmonary stenosis and anatomic contraindications to the Rastelli procedure, such as those with a small RV, remote VSD, or a straddling tricuspid valve, cone-shaped implants of the tricuspid valve, or the anterior implant of the mitral valve [61,62]. It consists in mobilizing the aortic root and coronary arteries posteriorly, doing a LeCompte maneuver, resecting the main PA and replacing it with a RV to PA conduit and closing the VSD. Ventosa-Fernández *et al.* [63] have reported that aortic translocation with the modified Nikaidoh procedure is a safe and effective surgical treatment for complex forms of TGA, particularly those associated with VSD and LVOTO. It is associated with lower reintervention and better morbidity and mortality. The short-term and mid-term survival up to 4.5 years has been 100%, and none of the patients required reintervention or mechanical circulatory support, when compared with the classical alternatives such as the Rastelli procedure.

Perioperative anesthetic management for arterial switch operation procedure

Preoperative evaluation and preparation

A detailed history and physical examination were required about gestational age, birth history, family history, and other associated medical problems. Moreover, assessment of the site for the arterial and intravenous cannulation was done, as these may be

difficult to insert and time consuming in some patients. The patient's hemodynamic data, oxygen saturation, vasoactive infusions, and medical or surgical interventions should be noted. A detailed information about the ventricular function, coronary arteries courses, and site and size of mixing between two parallel circulations should also be available. Laboratory parameters like complete blood count, platelet count, bleeding time, prothrombin time, serum electrolytes (sodium, calcium, and potassium), arterial blood gas, blood urea nitrogen, creatinine, and blood sugar should be available. A type and cross match should be sent to the blood bank to ensure the availability of adequate blood and blood products (FFP, platelets, and cryoprecipitate). Premedication is not necessary. All cardiac stabilizing medications (inotropes, prostaglandin, and pulmonary vasodilators) should be continued. The patients receiving PGE1 infusion to maintain or reopen the ductus arteriosus must be ensured that this is not discontinued at any stage during the transfer or induction of anesthesia. A subset of patients can require tracheal intubation and mechanical ventilation and subsequent emergency ASO because of cardiorespiratory compromise or episodes of cardiac arrest [64].

Anesthetic technique

Till date, there is no ideal anesthetic induction agent available without any cardiovascular effects in neonates and infants and neither any anesthetic technique has been shown to be beneficial. In practice, the relative advantage of a particular anesthetic technique is considered for the individual patient. The clinical spectrum of D-TGA is highly complex and variable, so it is not possible to be prescriptive. Most of the times, the anesthetic management is based on the institutional protocol and clinical experience with the anesthetic technique. All intravenous induction agents could be used at judicious doses [65,66].

Most of patients with D-TGA are shifted in the OR with intravenous access, so anesthesia can be safely induced and maintained using opioids in combination with inhalational agents (sevoflurane 2–3%) and benzodiazepine (midazolam 0.05–0.1 mg/kg). A narcotic-based technique using fentanyl (5–10 μg/kg) or sufentanil (10–15 μg/kg) or remifentanil (2–5 μg/kg) is safe as all are hemodynamically stable and blunt pulmonary reactivity, provide adequate analgesia, attenuate stress response, and ensure rapid and smooth induction. Fentanyl with a rapid onset and remarkable hemodynamic stability with a slight decrease in heart rate (HR) but no change in arterial pressure has been widely used in pediatric cardiac anesthesia. In addition, high doses (50–100 μg/kg) prevent rises in the plasma catecholamine and cortisol levels and prevent the stress response of the laryngoscopy, intubation, and sternotomy [67-69]. Sevoflurane (3–4%) is the preferred inhalational induction agent due to its rapid and smooth induction and very little myocardial depression and dysrhythmias. Sevoflurane up to 1–2 MAC maintains LV functions, without any significant decrease in HR, although systolic blood pressure decreases 20–25%. However, higher concentrations (8%) can lead to nodal rhythm and bradycardia, but cardiac index is well preserved [70]. Isoflurane is not suitable for induction of anesthesia in children due to propensity to cause coughing, breath holding, and laryngospasm [71]. However, it is used safely to supplement opioid anesthesia after induction to control hypertension and awareness. Isoflurane also preserves both CI and EF, is less arrhythmogenic, and produces lesser decrease in MAP as compared with halothane but increases HR [72]. It has been reported that all inhalational anesthetic agents not only cause myocardial preconditioning but also of brain, kidney, and other organs. The authors prefer judicious inhalational induction with sevoflurane (5–7%) till intravenous access is achieved, then sevoflurane is switched off, and fentanyl (5–10 μg/kg), thiopental (1 mg/kg), midazolam (0.05–0.1 mg/kg) and pancuronium (0.1–0.2 mg/kg) as muscle relaxants are administered. Perioperative hypertension at various stages can be managed with additional doses of fentanyl, and titrated doses of midazolam or sevoflurane/isoflurane. Some patients with inadequate blood mixing may become more hypoxic following induction. Here, a volume loading occasionally will improve the situation, and ensuring adequate depth of anesthesia may be helpful to decrease oxygen consumption.

Maintenance of preload, CO, HR, and decreasing PVR, and avoidance of myocardial depression and reduction in SVR are important hemodynamic goals, and to prevent further exaggeration in perioperative systemic arterial desaturation [73]. Low PVR in comparison with SVR ensures effective blood flow to lungs and thus intercirculatory mixing. In addition, increase in PVR may also diminish pulmonary blood flow from PDA or bronchopulmonary collaterals. The following measures can be used to ameliorate the rise in PVR and pulmonary hypertension: inhaled nitric oxide (NO), nebulized PGI2, inhaled milrinone, inhaled NTG and iloprost, and intravenous sildenafil, along with ventilatory interventions to increase FiO_2, avoid hypoxia, hypercarbia, acidosis, hypothermia, high and

low tidal volume, high PEEP and hypoglycemia, and light plane of anesthesia in neonates [72].

Intraoperative monitoring

Monitoring includes a five-lead electrocardiograph to detect any rhythm disturbances and myocardial ischemia. Dysrhythmias can result from several factors, including light anesthesia, hypoxemia, hypercarbia, drugs, electrolyte abnormalities, and various surgical maneuvers. The patient may become bradycardic or develop a nodal rhythm with induction or postrepair and often may require single or dual chamber pacing. Careful attention must be paid to the ST-segment patterns because myocardial ischemia can occur in these infants after ASO [74,75]. At this stage, nitroglycerin infusion (1–2 μg/kg/min) can aid in relaxation of the coronary vessels. Surgical revision of one of the coronary artery implantations may be necessary if there is too much stretch or kinking of the vessel. Other monitoring includes invasive arterial pressure (using umbilical, radial, femoral, or axillary artery), CVP, SpO_2, ABGs, Hct, ACT, serum glucose and potassium, urine output, and temperature. In the authors' institute, femoral arterial and right internal jugular venous pressure lines are preferred as these are more reliable and work for several days in the postoperative period. LA pressure monitoring is commonly used in patients undergoing ASO because of disparities in left-sided and right-sided heart function that are often present. The LA line is inserted by the surgeon at the end of repair. LA pressure should be maintained between 5 and 15 mmHg, with evidence of good tissue perfusion and urine output more than 1 ml/kg/h. It is recommended that milrinone (0.375–0.750 μg/kg/min) be started for any patient with signs or symptoms of low cardiac output and with at least a LA pressure more than 15 mmHg [26]. Equipment used to monitor central nervous system function may include electroencephalography, transcranial Doppler, jugular venous saturation monitoring, and near-infrared spectroscopy. Andropoulos *et al.* [66] have suggested the regional cerebral oxygen saturation monitoring throughout the perioperative period with a protocol that attempted to maintain regional cerebral oxygen saturation more than 50% before and after bypass and more than 90% while on bypass. TEE can also be used to assess the ventricular functions, evaluate surgical repair and volume status, and detect baffle leaks and residual defects. It can also detect the postrepair myocardial ischemia with the presence of RWMA and diastolic dysfunctions [76-78]. Attention should be paid to complications that could occur during the use of TEE, especially hemodynamic compromise from left atrial compression and VF due to probe in the small babies [26,79].

Cardiopulmonary bypass management

The general principles of CPB are same for neonates and adults. CPB requires aortic and bicaval cannulation, and rarely two aortic and three cava cannulas are essential in associated interrupted aortic arch and a persistent left superior vena cava for adequate drainage during the CPB. Use of large cannulas in a small-sized neonate can obstruct the venous and arterial flows before and after the bypass period. However, un-cannulated left superior vena cava can engorge sufficiently to cause cerebral ischemia. In addition, big major aorto-pulmonary collateral artery and PDA are concerns during CPB, as they can lead to hypoperfusion and wet lungs and need to be ligated before initiation of CPB. The hemodilution during CPB can decrease the hematocrit, coagulation factors levels (<30% of normal), platelet levels (<50 000/mm^3), and even the drug levels. Therefore, it is advised to keep the priming volume of CPB circuit to the minimum (95–110 ml) and may be primed with colloids (albumin) to avoid the hemodilution in neonates undergoing ASO [80]. Usually pump flow rates of 150–200 ml/kg/min are utilized, and mean arterial pressure of 30–35 mmHg, maintained with α-receptor blockade (phenoxybenzamine, phentolamine, or chlorpromazine). Hematocrit is maintained at 30–35% during cooling and hypothermic periods and at 40–45% during rewarming. Assessments of cardiac contractility (by direct visualization of heart and TEE), preload (by direct visualization of heart and via RA and LA pressures), PVR, SVR (by TEE), and HR should be done and should be optimized before coming off bypass. Milrinone (0.5 μg/kg/min), dobutamine (5 μg/kg/min), and nitroglycerin (1–2 μg/kg/min) infusions are often used to improve the LV contractility and augment CO and to reduce SVR and PVR. In addition, epinephrine (0.05–0.15 μg/kg/min) and norepinephrine (0.05–0.2 μg/kg/min) and calcium chloride (10 mg/kg) can also be added when necessary. In the authors' institute, a combination of dobutamine, milrinone, nitroglycerin, and calcium is preferred. In a subset of patients with unstable hemodynamics, epinephrine, levosimendan, and norepinephrine are also added. Post-bypass coagulopathy could be a particular problem and needs to be addressed [75]. Many approaches can be adopted to minimize transfusion requirements and preservation of platelet function and prevention of fibrinolysis like use of tranexamic acid – 10 mg/kg followed by 1 mg/kg hourly on CPB, or 50 mg/kg of

epsilon aminocaproic acid followed by infusion of 25 mg/kg/h or aprotinin – 60 000 kallekrein inhibiting units (KIU)/kg loading dose followed by infusion of 7000 KIU/kg/h, with bypass circuit prime of 60 000 KIU/kg, and recombinant factor VII (30–90 μg/kg) in cases with severe bleeding [26,75,79,81]. Some authors have proposed the use of fresh blood up to 5 days old, because compared with stored RBCs, fresh RBCs are more metabolically balanced, have a higher pH, contain less potassium and lower concentrations of lactate, and cause lower pulmonary complications, renal dysfunction, and infection rates [82]. It has been observed that perioperative hyperglycemia and mild transient hypoglycemia do not appear to have any adverse outcomes in the infants with congenital heart disease [83].

Usually, conventional ultrafiltration is used throughout the bypass period to decrease tissue edema and inflammatory mediators. In addition, the modified ultrafiltration (MUF) can also be utilized selectively in the immediate post-bypass period. Here, the aortic cannula is left in place, and ~10–30 ml/kg/min of blood is siphoned from the aorta, pumped through a hemoconcentrator (dialysis membrane), and returned to the right atrium. It ameliorates many of the adverse effects of CPB and improves the outcome through various effects: improves hemodynamics; reduces total body water; decreases the need for blood transfusions; increases hematocrit, fibrinogen, and total plasma protein levels; improves intrinsic LV systolic function and diastolic compliance; increases blood pressure; decreases inotropic requirement in the early postoperative period; and decreases duration of mechanical ventilatory, length of ICU stay, and early morbidity and mortality [84-87]. Mahmoud *et al*. [88] have reported that MUF significantly decreases levels of the lung vasoconstrictor endothelin-1 and pulmonary-to-systemic pressure ratio after CPB and prevents pulmonary hypertensive crises (PHC) after pediatric cardiac surgery. It improves lung compliance, reduces cytokine levels, and removes activated complement (C3a and C5a) [89].

Postoperative management

Low cardiac output syndrome (LCOS) following ASO is the prime concerned and multifactorial factor that is related to CPB and circulatory arrest or to myocardial preservation, mechanical disruption of the myocardium, post-inflammatory effects of bypass, coronary manipulation, secondary to anomalous coronary artery patterns, and 'unprepared' LV. The LCOS is considered if patients met more than two of following diagnostic criteria: CI less than 2 l/min/m^2, systolic blood pressure decreased by more than 20% compared with preoperative blood pressure, CVP more than 15 cmH_2O, difference between the central temperature and the peripheral temperature more than 5°C, and the cold limbs, oliguria (diuresis <0.5 ml/kg/h), central venous oxygen saturation less than 60% (with normal arterial oxygen saturation) and/or lactate more than 3 mmol/l, without relative hypovolemia, elevated LAP, and decreased cerebral NIRS [89-91]. LCOS needs to be addressed promptly with pharmacological support using vasoactive, inotropic, and lusitropic drugs (levosimendan, milrinone, dobutamine, and epinephrine) [90]. Any suspicion of coronary insufficiency should be addressed immediately with possible re-exploration and revision. Li Xing Zhu *et al*. [64] and Hong *et al*. [92] have presented trials on patients with TGA undergoing ASO and described the early postoperative complications like delayed sternal closure (3.63±1.49 days), prolonged mechanical ventilation time (5.89±3.02 days) and an ICU stay (10.12±3.25 days), and infection. One of the most common arrhythmias after an ASO is junctional ectopic tachycardia. Often the management strategies for JET include adequate sedation, controlled hypothermia (35°C–36°C rectal), decreasing of vasoactive agents doses, optimization of electrolytes, use of atrial pacing over ventricular rate controlled for brief period. Following these the rhythm should be reassessed and if it is unresolved then the medications like; Dexmedetomidine (0.2 mcg/kg/hr up to max of 1 mcg/kg/hr), amiodarone(1 mg/kg over 10 minutes, repeat in 1mg/kg increments to a total dose of 5 mg/kg), procainamide (7–10 mg/kg load over 30 minutes) and then infusion at 40–60 mcg/kg/min, and esmolol (50–300 mcg/kg/min) may be considered. Other antiarrhythmic drugs commonly used in the neonate after an ASO are sotalol, propranolol, digoxin, adenosine and magnesium sulphate [26]. In patients with TGA/VSD, the evaluation of pulmonary hypertension is very important to avoid postoperative fatal PHC causing RV failure [64]. The medical treatments for PHC should be initiated immediately with inhaled nitric oxide, and other intravenous and inhaled pulmonary vasodilators (milrinone, prostaglandins, and NTG). These patients can suffer from persistent bleeding and tamponade during the early postoperative period and require emergent mediastinal exploration in the ICU. Ventilator-associated pneumonia may also occur following mechanical ventilation. Central nervous system complications like intraventricular and intracranial hemorrhage with or without hydrocephalus may present with seizures and require anti-epileptic

medication. Some patients may develop acute renal failure and require hemodialysis assistance [93].

Mortality occurs owing to severe LCOS, PHC, multiple organ dysfunction, convulsions, and refractory ventricular arrhythmias. Some authors have described the mortality risk predictors like low APGAR-score, older age at surgery, necessity of associated surgical procedures, lesser institutional and surgeon experience, smaller patient size, side-by-side great vessel arrangement, LV hypoplasia, LV outlet obstruction, and arch abnormalities, as well as intramural and single coronary artery. [1,94,95].

Conclusion

Nowadays, the TGA can be diagnosed accurately in the prenatal period by screening the fetal heart at the time of the obstetric anomaly scan (18–22 weeks of gestation), and the type of repair likely to be required after birth can also be well predicted beforehand. The initial management includes PGE1 infusion, BAS, iNO, vasoactive agents, ventilatory support, or ductus stenting and RVOT dilatation. Currently, the ASO is the gold standard operative correction of neonates with TGA with excellent operative survival; 30-day mortality rate for the ASO at less than 3% with a 1-year survival rate of more than 96% [1,10]. ASO can be performed safely under narcotic-based balanced anesthesia technique using fentanyl, sevoflurane, and midazolam, as it is hemodynamically stable, blunts pulmonary reactivity, provides adequate analgesia, attenuates stress response, and also ensures rapid and smooth induction. Maintenance of preload, CO, and HR; decreasing PVR; avoidance of myocardial depression; and reduction in SVR are important hemodynamic goals during perioperative management. Some authors have suggested the selective use of MUF following ASO repair to ameliorate the complications of the prolonged CPB provides early outcome benefits [86-90]. Postoperatively, these patients may suffer from LCOS, myocardial dysfunctions due to coronary issues and prolonged duration of CPB, arrhythmias, PHC, extensive bleeding, and cardiac tamponade; therefore, optimizing postoperative treatment helps to improve the survival rate after ASO. ECMO is vital in extreme cases for hemodynamic and respiratory support, bridge to ASO or even for patients with CPB weaning failure, and postoperative cardiorespiratory support for refractory LCOS, PHC, RV failure, and unprepared LV.

Conflicts of interest

There are no conflicts of interest.

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