

Transposition of the Great Arteries

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Transposition of the great arteries (TGA, simple transposition of the great arteries, or D-transposition of the great arteries) is a common cause of prostaglandin-dependent cyanosis and the most common cause of cyanosis in the newborn. It occurs when there is ventriculoarterial discordance and there is recirculation of pulmonary and systemic blood flows. The most common anatomical presentations are with an intact ventricular septum (IVS), a VSD, and VSD with LVOT obstruction (LVOTO) (Figure 14-1). In the absence of intracardiac mixing (PDA, ASD, or VSD), it is rapidly fatal. In the current era, expectations for effective treatment (neonatal surgical repair) should be very high.

Pathophysiology and Clinical Presentation

During fetal life, oxygenated blood from the placenta will cross the PFO and through the main PA and PDA, perfuse the distal aorta. The highly deoxygenated blood from the SVC that enters the RV through the tricuspid valve will perfuse (through the aortic valve) the aortic arch and neck vessels.

After birth, circulation will be in parallel (Figure 14-2). While patent, the arterial duct promotes the pressure differential to enhance atrial mixing by increasing LAP. When there is a marked difference (>5%) in upper/lower extremity saturations and postductal saturations are higher than preductal, the clinician should suspect aortic arch obstruction. When the atrial communication is small or restrictive, infants will be profoundly desaturated (oxygen saturation in the 60s). Given that atrial mixing takes place due to the pressure differential between both ventricles in diastole, it is possible that even in the presence of a large unrestrictive ASD, mixing could be insufficient. A large unrestrictive VSD may be an effective place for mixing when is part of the presenting anatomy, but smaller defects may also be insufficient.

On physical exam, the child will become tachycardic and tachypneic. Grunting can be present when there is acidosis and/or pulmonary edema. The precordium is hyperactive with an RV impulse. The second heart sound will be single from elevated PA pressures.

Diagnosis

- **CXR.** Usually without cardiomegaly immediately after birth. The upper mediastinum may be narrow due to the anteroposterior relationship of the great vessels (“egg-on-a-string” sign).
- **ECG.** Usually normal.
- **Echocardiogram (Figure 14-3).** Echocardiogram is the cornerstone of diagnosis. On the parasternal long-axis view, the great arteries will appear in parallel with the great artery arising from the LV (PA) taking a posterior turn (pathognomonic). One should then detour from the standard echocardiogram protocol to switch to coronal subcostal imaging and demonstrate the atrial septal communication. Thereafter, the focus should be on the arterial duct and its patency. This information

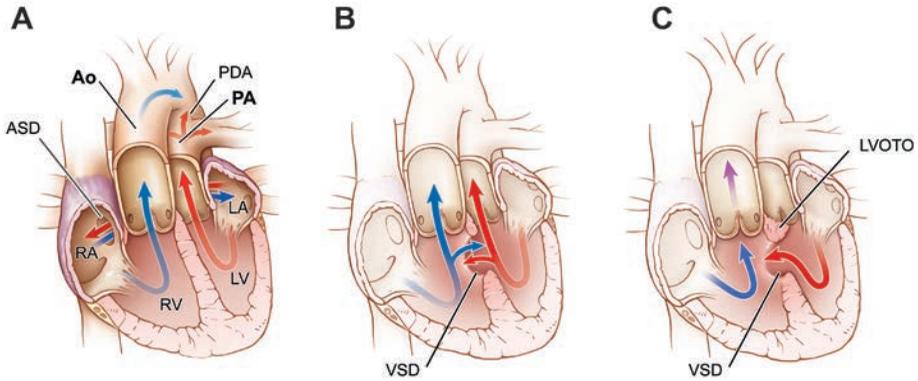


Figure 14-1. Anatomical variants of TGA. A: TGA with IVS. B: TGA with VSD. C: TGA with VSD and LVOTO. Ao: Aorta.

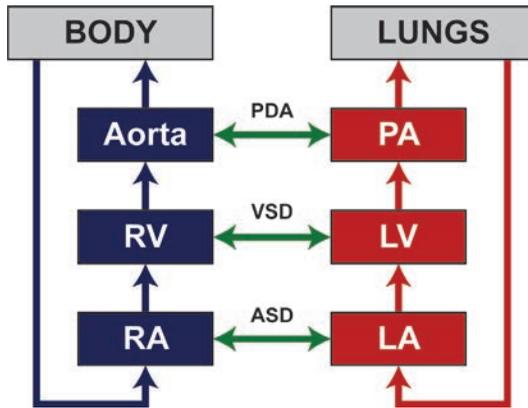


Figure 14-2. Physiology in TGA. The systemic and pulmonary circulations run in parallel, relying on the presence of patent communications (PDA, ASD, or VSD).

is the most relevant when trying to establish the initial diagnosis and to mobilize the interventional cardiology staff for a potential bedside balloon atrial septostomy (BAS). As usual, the echocardiogram laboratory protocol should be completed before making final determinations about interventions. The coronary arteries should be resolved by echocardiogram. The most common coronary artery patterns are the following: right and left coronaries (including LAD and circumflex) from usual sinuses (Yacoub A), left circumflex origin from the right coronary artery (Yacoub D), and single origin of the right or left systems (Yacoub B) (Figure 14-4).

- **Cardiac CT.** Usually not necessary. Postoperatively, if there is a concern about the patency of the coronary arteries, the child should undergo cardiac catheterization or return to the OR without delay.

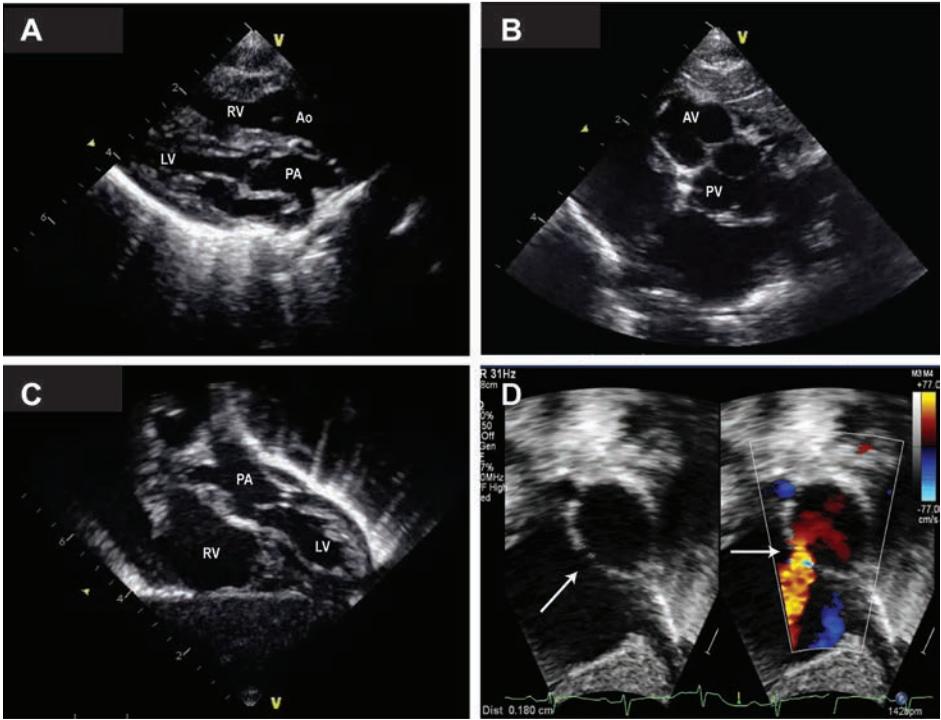


Figure 14-3. Echocardiography in TGA. A: Parasternal long-axis view showing the PA originating from the LV and the aorta (Ao) from the RV. B: Parasternal short-axis view showing the aortic valve (AV) located anterior and rightward to the pulmonary valve (PV). C: Subcostal view again showing the PA originating from the LV. D: Subcostal color-compare image depicting a restrictive atrial septum (arrow). Images courtesy of Dr. Josh Kailin, www.pedecho.org.

- **Cardiac catheterization.** Although the hemodynamic definition of TGA is when the saturation in the PA is higher than the aorta, cardiac catheterization is seldom necessary for diagnosis. After birth, most children will undergo BAS to improve mixing at the atrial level (unless the defect is large and unrestrictive). If there are concerns/questions about coronary artery anatomy, an extreme caudal down-to-barrel angiogram may be performed, although in the current era, precise delineation of the coronary anatomy is not necessary prior to surgery.

Indications / Timing for Intervention

A BAS should be performed in most patients to allow adequate mixing and stabilization of the infant, unless the defect is large and unrestrictive. It is generally performed at the bedside or cardiac catheterization laboratory with either a Rashkind or a Braun catheter. There should be a visible tear on the septum primum allowing for bilateral motion of the remaining flap by echocardiogram and an improvement in oxygen saturations.

The arterial switch operation (ASO) is usually performed after the PVR has decreased

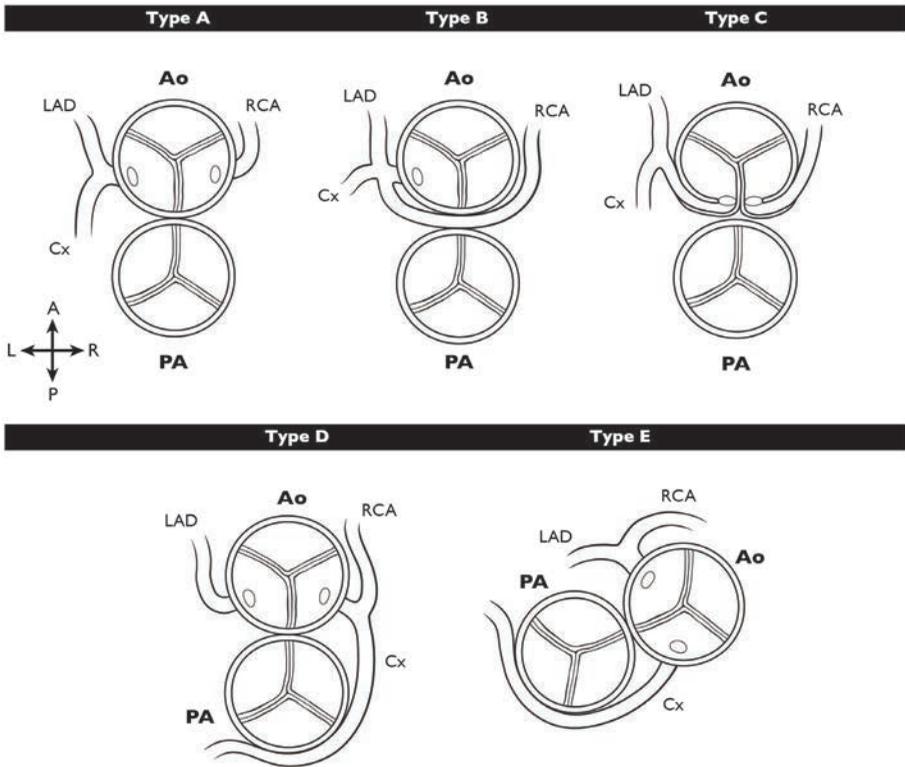


Figure 14-4. Common coronary patterns in TGA (Yacoub and Radley-Smith classification).

(>48 hrs), although a rare patient may present with intractable cyanosis requiring a more urgent ASO. Children with a restrictive ASD/PFO have a slower transition of the PVR and may present with persistent cyanosis even after the BAS. Timing of the ASO is usually on the first week of life. In patients with adequate saturations and non restrictive VSDs, the morphologic LV will not decondition and the ASO may be deferred to a semielective status but within the same hospital admission.

Anesthetic Considerations

Typical anesthetic agents for ASO include fentanyl at 25-100 mcg/kg total dose, isoflurane at any required dose, low-dose midazolam at 0.2-1 mg/kg total dose, and dexmedetomidine, either started before incision with a loading dose, or after aortic cross-clamping, at reduced doses of 0.2-0.5 mcg/kg/hour because of reduced clearance in the neonate.

The anesthetic approach will depend on whether there is adequate mixing (i.e., BAS performed with large ASD, PDA with prostaglandin still infusing, or VSD). If there is significant oxygen desaturation (i.e., $\text{SaO}_2 < 80\%$) at baseline, there is a significant risk of further desaturation, lower cardiac output, and decreased mixing with the induction

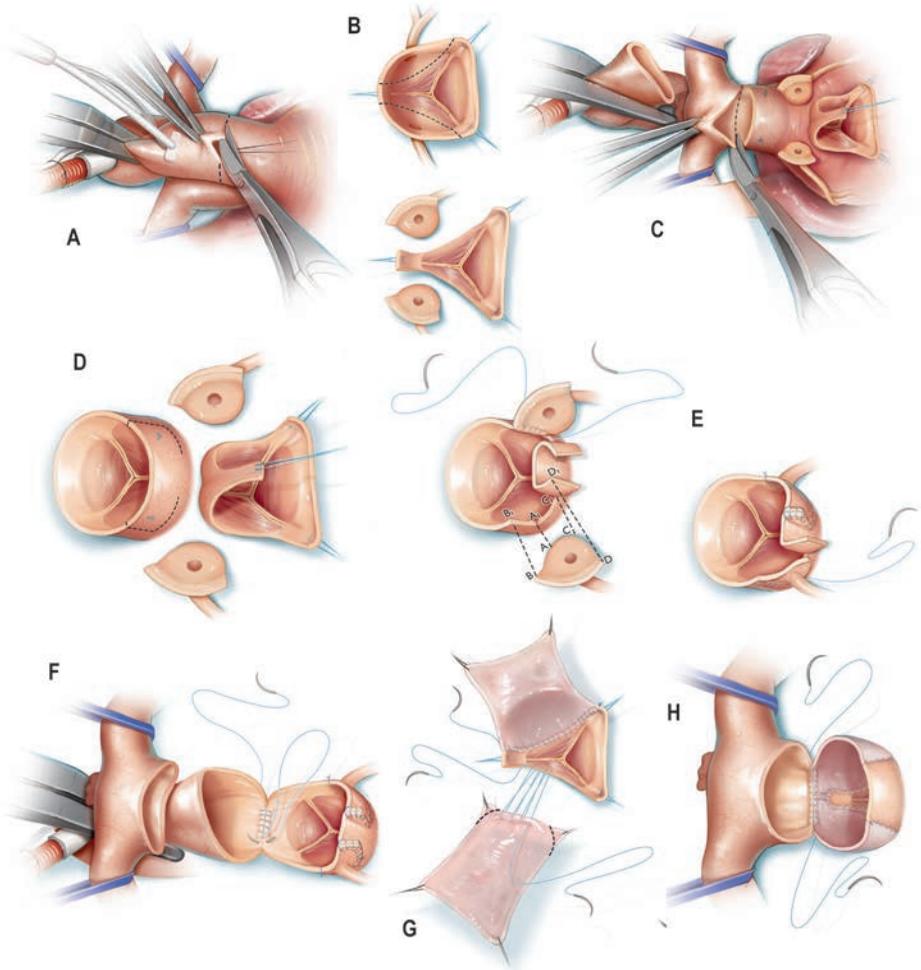


Figure 14-5. Arterial switch operation. See text for details.

of anesthesia. If this is the case, increasing cardiac output with inotropic support (epinephrine 0.02-0.03 mcg/kg/min), increasing FiO_2 to 1.0, and increasing hemoglobin to 13 g/dL or higher with PRBC transfusion is usually effective. Patients with multiple sources of mixing and $\text{SaO}_2 > 90\%$ will often require low FiO_2 of 0.21-0.3 before CPB, and other measures to increase PVR, such as mild hypercarbia and positive end-expiratory pressure of 5-8 cm H_2O . Establishing a baseline cerebral rSO_2 with baseline conditions is important, and treating $\text{rSO}_2 < 50\%$ absolute values, or $> 20\%$ relative decrease from baseline is an effective approach.

During CPB for the arterial switch operation, the anesthesiologist works closely with the perfusionist and surgeon to optimize conditions of oxygen delivery to the

brain and other vital organs. Flow rates of 150 mL/kg/min are normally used, with CPB MAPs usually around 40 mmHg for neonates. Phentolamine at 0.05-0.15 mg/kg in divided doses is often used to maintain high flows with low perfusion pressures. rSO_2 is monitored bilaterally and pH-stat CPB management used, with hematocrit goals at about 30%, to maintain rSO_2 well above baseline during CPB. Weaning from CPB is accomplished gradually using an LA catheter for guidance; LAP is kept at 0-5 mmHg during the weaning process, with a usual goal of 4-8 mmHg immediately after CPB. LV distention (LAP >10 mmHg), particularly in the patient with preoperative IVS, is to be assiduously avoided, because excessive preload will not be tolerated. If during the weaning process LAP is >10 mmHg, intravascular volume is taken from the patient, either into the CPB circuit, or by removal of blood by the surgeon or anesthesiologist.

After CPB, low-dose epinephrine (0.02-0.05 mcg/kg/min), nitroglycerine at 1 mcg/kg/min, and calcium chloride infusion at 5-10 mg/kg/hr are standard infusions. Vasopressin at 0.02-0.04 units/kg/hour can be utilized to increase perfusion pressure, and sodium nitroprusside at 0.5-2 mcg/kg/min can be utilized to decrease BP. Pressure-controlled ventilation with FiO_2 1.0 is utilized to wean from CPB, and FiO_2 can later be reduced, if appropriate.

TEE is often utilized to assess for any residual defects such as VSD, and for myocardial function and sequential wall motion abnormalities of the LV. However, in smaller neonates, TEE may not always be used and clinical status, ST-segment changes, and appearance of the heart are assessed to judge adequacy of repair. In those cases, an epicardial echocardiogram may be utilized if there are questions about anatomy or myocardial function.

The goals of low LAP, MAP in 40s-50s, normocarbia, and maintaining adequate cardiac output are meticulously pursued. After protamine administration, platelet infusion of 10-15 mL/kg is often sufficient to decrease bleeding. Cryoprecipitate at 5-10 mL/kg is the next choice of coagulation products, followed by FFP 10-20 mL/kg. Coagulation product administration can be guided by ROTEM® during rewarming on CPB. If bleeding continues after ruling out surgical causes and administration of 2 or more doses of platelets, cryoprecipitate, and FFP, activated factor VII in doses of 45-90 mcg/kg can be considered after thorough discussion. Although there is theoretical risk of thrombosis from this agent, in practice this has not been observed. The activated factor VII dose can be repeated in ~90-120 minutes if bleeding is ongoing.

Surgical Repair

Anatomic repair (ASO) (Figure 14-5) is now the standard of care for treatment of the majority of TGA patients with IVS, VSD, and aortic arch obstruction (in addition to patients with Taussig-Bing anomaly, see Chapter 16). In developed countries, it is now very unusual for a baby with TGA to present beyond the newborn period for primary treatment, however in some areas of the world, this is more common. In patients presenting with TGA/IVS beyond 6 weeks of life, the LV may have involuted and thereby will not be capable of managing a systemic workload immediately after the ASO. One option is to place a PA band (PAB) to rapidly retrain the LV (in small infants, this usually can happen in a period of approximately 1 week but will frequently require a systemic-to-PA

shunt to provide adequate pulmonary blood flow), followed by an ASO. Retraining the morphologic LV, however, is not only time-consuming, but is a risk-laden proposition. As such, some centers recommend an atrial switch operation (Senning or Mustard) for the late-presenting patient with TGA/IVS. Yet another option is to proceed with an ASO in the first 3 months of life with the understanding that temporary mechanical assistance (VAD) may be needed postoperatively.

For the typical newborn patient with TGA/IVS, the ASO is performed within the first week of life, although we have successfully performed a primary ASO up to 8 weeks of life with good results. The ASO is performed via median sternotomy on CPB support. We have favored separate caval venous cannulation in all except the very smallest of babies (<2 kg) along with a single aortic cannula placed in the distal ascending aorta (in patients with severe aortic arch obstruction, a second arterial cannula in the ductus may be needed for perfusion of the lower body). In TGA/IVS, mild hypothermia (nasopharyngeal temperature of $\sim 32^{\circ}\text{C}$) as an adjunct to myocardial and brain preservation. As noted previously (see Chapter 6), we favor a high-flow, low-pressure perfusion strategy individualized to the patient's needs.

During preliminary dissection, fresh, autologous pericardial patches are harvested and prepared for later use in reconstructing the neo-pulmonary sinuses of Valsalva. The coronary ostial locations are noted and marking sutures are placed on the pulmonary root (neo-aortic root) to help with coronary ostial translocation. This coronary movement is the key element of the ASO and maneuvers have been well-described to facilitate accurate translocation of all coronary branching patterns including single coronary ostium and intramural coronaries. The surgeon must be prepared for all contingencies.

After the establishment of safe CPB, the patient is gradually cooled. During this phase, the ductus is ligated and divided and the branch PAs are widely mobilized to facilitate anterior positioning of the PAs after reconstruction (maneuver of LeCompte). Following standard cardioplegic arrest, the ascending aorta is transected just at or above the sinotubular junction (mindful of anomalous aortic origins or intramural coronaries) and the ostia inspected (Figure 14-5, A). The coronary ostia are then mobilized as liberal buttons of aortic wall (Figure 14-5, B) and the proximal coronaries are also mobilized with great care not to skeletonize the actual coronary artery. Next, the main PA is transected (also just at or above the sinotubular junction) (Figure 14-5, C). For the majority of patients, we create appropriate, medially based, trapdoor flap incisions in the neo-aortic sinuses to facilitate ostial translocation with minimal axial rotation (the key concern in problematic ostial transfer) (Figure 14-5, D). The coronary buttons are anastomosed using very fine monofilament suture (7-0 or 8-0 polypropylene) (Figure 14-5, E). After the maneuver of LeCompte, aortic continuity is reestablished by a primary anastomosis between the ascending aorta and neo-aortic root (Figure 14-5, F). As this is occurring, gradual warming of the patient is commenced and the heart is vigorously deaired. The ASD is then completely closed (typically can be done primarily although in some cases, a patch may be needed). The heart is vented through the anterior ascending aorta and reperfused. A normal sinus rhythm should resume spontaneously and the ECG should rapidly normalize. Persistent ST-segment changes may be related to intracoronary air or the more concerning possibility of coronary ostial malpositioning.

This latter concern is critical and must be assessed at this point. Once the surgeon is satisfied with the coronary translocation, the neopulmonary sinuses are individually reconstructed (deficiencies that were created by the coronary ostial mobilization) with liberal patches of the previously harvested, fresh autologous pericardium (Figure 14-5, G). Finally, PA continuity is reestablished by a primary anastomosis between the neopulmonary root and PA bifurcation (Figure 14-5, H). In patients with side-to-side great vessel relationship (such as Taussig-Bing anomaly), the neopulmonary-to-main-PA anastomosis may need to be placed out onto the right PA to prevent distortion or compression of the translocated coronary buttons.

In cases where there is a VSD, it is our practice to proceed with VSD closure prior to the ASO. There are several reasons for this sequence. First, in cases with a degree of malalignment of the great vessels to the ventricular septum (very important in cases of Taussig-Bing anomaly, it is critical for the surgeon to be certain that the VSD closure can be committed to one or the other great vessel. Secondly, it is best not to place traction on the reconstructed great vessels after the ASO. Another concern may be the status of the semilunar valves or LVOT, which may often be assessable through the VSD before committing to the ASO. We favor autologous pericardium for VSD closure(s). It is not uncommon for us to reperfuse the heart (by removing the cross-clamp while making the tricuspid valve incompetent and venting the aortic root) for 10-15 minutes between VSD closure and the ASO in order to limit continuous cross-clamp time.

In cases of aortic arch obstruction, the patient will be cooled to a more profound level (nasopharyngeal temperature ~18-20 °C) and then repair the arch primarily (see Chapter 25). Following arch reconstruction, the patient is partially rewarmed while the VSD is closed and the ASO performed.

In patients with significant LVOTO, the ASO may still be possible if the subaortic area is amenable to resection. In patients with congenitally bicuspid pulmonary (neoaortic) valves, the ASO is still feasible, assuming the valve caliber is adequate.

In patients with Taussig-Bing anomaly, there may be an enormous size discrepancy between the aortic root and the pulmonary root (with the pulmonary root being much larger). In this setting, reconstruction of the ascending aorta may require augmentation for an effective anastomosis. Since these patients also often have actual or impending RVOT obstruction, it is often wise to perform a prophylactic RVOT resection prior to reconstruction of the neopulmonary root.

Prior to weaning from CPB, an LA catheter is placed. Information about the LAP

ASO experience (<18 months old) at TCH (1995-2018)

Anatomy:

- TGA/IVS: 204 (52%)
- TGA/VSD: 137 (35%)
- TGA/VSD/LVOTO: 17 (4%)
- Taussig-Bing anomaly: 36 (9%)

Median age at surgery: 8 days (1 day – 17 months)

Median ICU length of stay: 6 days (5-8 days)

Median hospital length of stay: 11 days (9-16 days)

Perioperative mortality: 1.3%

5-year survival: 98.2%, 10-year survival: 97.8%, 15-year survival: 97.8%

Postoperative mechanical circulatory support / ECMO: 2%

is critical to individualized patient management. Intraoperative assessment of the coronary arteries by transesophageal or epicardial echocardiography is obtained to confirm adequate coronary blood flow. A peritoneal dialysis catheter is routinely placed. Hemostasis must be achieved prior to leaving the OR. It is very unusual to need to leave the sternum open.

Postoperative Management

The chief concerns on the postoperative period include the effectiveness of coronary blood flow and the ability of the LV to accommodate to systemic workload. Children with a restrictive ASD preoperatively will be at higher risk of persistently higher PVR and in rare circumstances may require iNO in the early postoperative period.

General Management

- **Fluids.** 25% maintenance with D5%/0.45%NS is standard. Careful attention should be paid to managing the patient with the minimal necessary preload. Unnecessary preload increases may produce increases in myocardial wall stress and lead to ventricular dysfunction and hypotension. This may occur with very small volumes of excess fluid administration (as small as <5 mL total in a single bolus) and is another reason to emphasize the proper use of LAP in perioperative management.
- **Analgesia and sedation.** Analgesics and sedatives are adjusted for patient's comfort. Fentanyl (1-3 mcg/kg/hr) infusion is commonly used for analgesia. *Sedation* is achieved with a combination of dexmedetomidine (both intubated and extubated patients) and/or benzodiazepines. Midazolam as a drip is preferred, as significant shifts in afterload or BP may produce instability.
- **Vasoactive drugs.** Most patients will arrive from the OR on milrinone (0.25-0.75 mcg/kg/min) and sometimes epinephrine (0.02-0.05 mcg/kg/min). Hypotension should be primarily managed with inotropes when LV filling pressure (LAP) is higher than 5-10 mmHg.
- **Mechanical ventilation.** Patients are usually ventilated on SIMV-VC with pressure support, at Vt of 8-10 ml/kg and PEEP of 5-7 mmHg, and aiming for pH 7.35-7.45 and SaO₂ >95%. After the ASO, patients may be extubated in the first postoperative day if hemodynamics are adequate.

What to Expect in the First 24 Hours Postoperatively

- **Vasoactive drugs.** It is reasonable to manage milrinone and low-dose epinephrine (<0.03 mcg/kg/min) through extubation to support the LV, as extubation will lead to high SVR, increase in transmural pressure, and consequently higher afterload.
- **Ventilation.** Transitioning from the OR, the lungs will be significantly improved from the preoperative period secondary to continuous ultrafiltration.
- **Fluids.** Even to slightly negative. The peritoneal dialysis catheter should be used starting on the day of surgery.
- **Nutrition.** If considering extubation within 24 hrs, it is not necessary to write for TPN. If longer periods of mechanical ventilation are anticipated, full TPN should be ordered. Oral feeds should be reinstated once successfully extubated.

Complications

The most common post-operative complications after TGA repair are LCOS and cardiac arrhythmias.

- **Coronary artery issues.** Although all coronary artery patterns are able to be managed with the ASO, several multicenter studies have shown that there is increased mortality in complex patterns, in particular intramural coronaries. A low index of suspicion is necessary as coronary issues may present with increased LAP and LCOS, without clear ECG changes.
- **LCOS** (see Chapter 71). Primarily treated with inotropes. A combination of low-dose epinephrine and standard-dose milrinone. High inotrope doses increase the likelihood of arrhythmias. If prolonged, severe, or associated with significant volume requirements, it should prompt reopening of the sternum, visual and echocardiographic inspection of the coronary arteries and either return to the OR or coronary artery evaluation by cardiac catheterization.
- **Arrhythmias.** Arrhythmias or heart block are unusual. Persistent atrial tachycardia or JET (see Chapter 74) may decrease cardiac output.
- **Bleeding** (see Chapter 76).
- **Prolonged mechanical ventilation.**
- **Chylothorax** (See Chapter 77). Potentially from high RV pressures, pulmonary hypertension, or increased tricuspid valve regurgitation in the presence of branch PA stenosis.
- **Mechanical circulatory support.** Any need for mechanical circulatory support after the ASO should prompt cardiac catheterization and coronary artery evaluation. In patients presenting for a primary ASO after 6 weeks of life in the setting of TGA/IVS, one should be prepared for temporary LV mechanical support in the acute perioperative period.

Long-Term Follow-Up

Despite excellent LV recovery and long-term survival rates after TGA repair, surveillance for ongoing complications including aortic root dilation, coronary insufficiency, and branch PA stenosis are necessary for life. In addition, prolonged deep-hypothermic circulatory arrest time, prematurity, and associated genetic syndromes are risk factors for suboptimal short- and long-term neurodevelopmental outcomes.

Suggested Reading

Dibardino DJ, Allison AE, Vaughn WK, et al. Current expectations for newborns undergoing the arterial switch operation. *Ann Surg* 2004;239:588-596.