28 Truncus Arteriosus and Aortopulmonary Window

Antonio G. Cabrera, Stuart R. Hall, Carlos M. Mery

Truncus arteriousus is a rare congenital heart anomaly comprised of a single origin of 3 vascular structures: the aorta, the PAs, and the coronary arteries (Figure 28-1). An aortopulmonary (AP) window is a proximal communication between the aorta and PA, with separate origins of both vessels, due to an incomplete division of the common arterial trunk (Figure 28-2). Even though both lesions share some common characteristics, in truncus arteriosus there is only one semilunar valve and one arterial trunk, contrary to an AP window, where there are two. The truncal valve is usually tricuspid, although it may also be quadricuspid or bicuspid.

As is the case with other conotruncal abnormalities, truncus arteriosus is associated with DiGeorge syndrome. AP window may also be associated with other extracardiac anomalies such as the VACTERL association (vertebral anomalies, imperforate anus, cardiac anomalies, tracheoesophageal fistula, renal anomalies, and limb abnormalities).

Classification

The most commonly used classification of truncus arteriosus is the classification of Collet and Edwards (Figure 28-1):

- **I:** A single PA arises from the common trunk and divides into 2 PA branches
- **II:** Two separate posterior or posterolateral PA branches arise from the common trunk
- **III:** Two lateral PA branches arise from the common trunk
- **IV:** The PA branches arise from the descending aorta (hemitruncus). This class is no longer used as it is more appropriately classified as pulmonary atresia with VSD and major aortopulmonary collaterals (see Chapter 17).
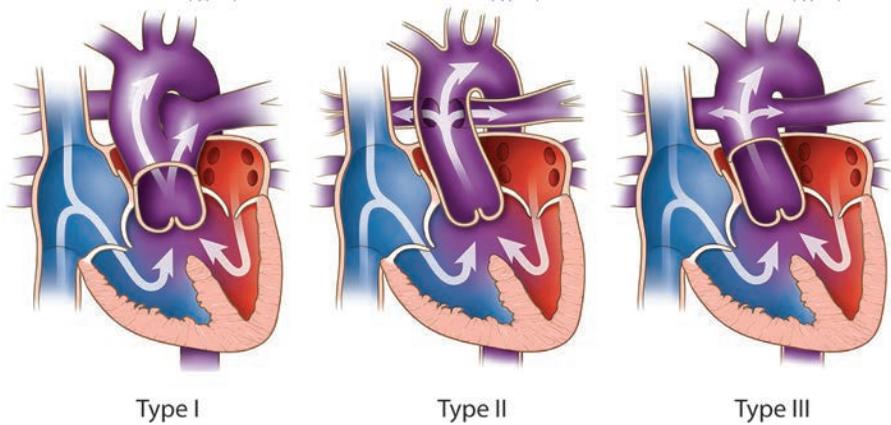
Most patients with truncus arteriosus present with what is called "type one-and-a-half" since both branch PAs arise very close to each other.

An alternative classification is the Van Praagh classification in which A1 is equivalent to type I, A2 combines types II and III, A3 describes a single origin of a PA from the truncus with a collateral or PDA perfusing the other PA, and A4 is truncus arteriosus with an interrupted aortic arch.

A more recent classification by the STS divides truncus it into either aortic or pulmonary dominance. Most cases are characterized by aortic dominance with a large ascending aorta and aortic arch, and smaller PA branches arising from the trunk. In pulmonary-dominant truncus, the ascending aorta is hypoplastic and the descending aorta is supplied by the PDA. Truncus arteriosus with interrupted aortic arch is an example of pulmonary dominance.

Pathophysiology and Clinical Presentation

Both truncus arteriosus and AP window are characterized by a significant left-to-right shunt resulting in overcirculation. The degree of overcirculation in patients with an AP window is obviously dependent on the size of the defect.

Type I   Type II   Type III

Figure 28-1. Collet and Edwards classification of truncus arteriosus (see text for details).

Patients with truncus arteriosus or a large AP window will present with tachypnea and pulmonary edema shortly after birth. The pulmonary overcirculation and CHF symptoms worsen as the PVR decreases with time. If the lesion is associated with aortic arch obstruction, shock and cyanosis may be presenting signs. Patients with a small AP window may present later in life and this lesion may be missed in the presence of other lesions such as tetralogy of Fallot, VSDs, and PDA.

If the lesion is not repaired, PVR will slowly increase and lead to irreversible pulmonary vascular changes (Eisenmenger syndrome). As a result, the shunt will decrease with time and once the PVR is significantly higher, it will reverse and become a right-to-left shunt with resulting cyanosis.

On physical exam, the child will be tachycardic and tachypneic. Grunting can be present due to pulmonary circulation and pulmonary edema. The precordium is hyperactive. Patients with truncus arteriosus will have a single S_2 and a systolic ejection click. Patients with an AP window will have a continuous mid frequency murmur, similar to a PDA. When presenting late, these patients may present with a single S_2 due to elevated PA pressure and early P_2 closure.

Diagnosis
- **CXR (Figure 28-3).** Cardiomegaly with pulmonary edema and pulmonary hyperinflation.
- **ECG.** Right-axis deviation. There can be biventricular hypertrophy with non-specific ST-T wave changes. In cases of coronary artery insufficiency, ST-segment depression may be present.
- **Echocardiogram (Figure 28-4).** Important features to assess in patients with truncus arteriosus include the truncal valve, VSD, coronary arteries, and associated anomalies, including the aortic arch. Because the combined ventricular output is going through the single truncal valve, there will be increased velocity across
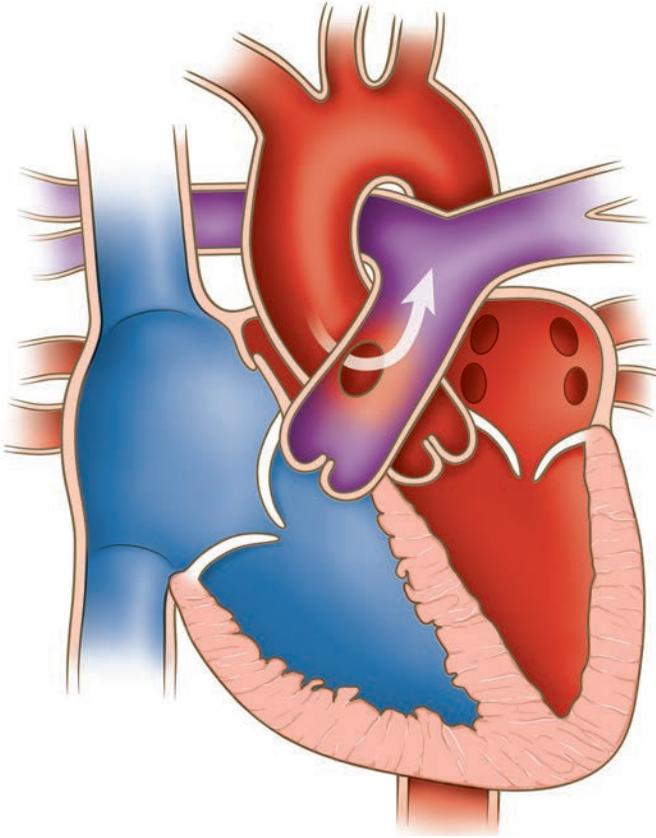
Figure 28-2. AP window.

it. Velocities higher than 3-3.5 m/s should increase suspicion for truncal valve stenosis. Imaging in patients with AP window should be focused on the location of the window and associated cardiac defects, such as tetralogy of Fallot, subaortic stenosis, and PDA.

- **CTA.** Usually not necessary for patients with truncus arteriosus. However, it can be useful if the anatomy is not completely resolved with echocardiogram. It may also be useful for patients with AP window to assess the location and extent of the window, and help with surgical planning.
- **Cardiac catheterization.** Not usually necessary for diagnosis except in cases with late presentation to evaluate PVR and operability.

Indications / Timing of Intervention

A diagnosis of truncus arteriosus or AP window is an indication for surgical intervention. The prognosis of patients with unrepaired truncus arteriosus is dismal due to the severity of CHF symptoms and the rapid development of pulmonary vascular
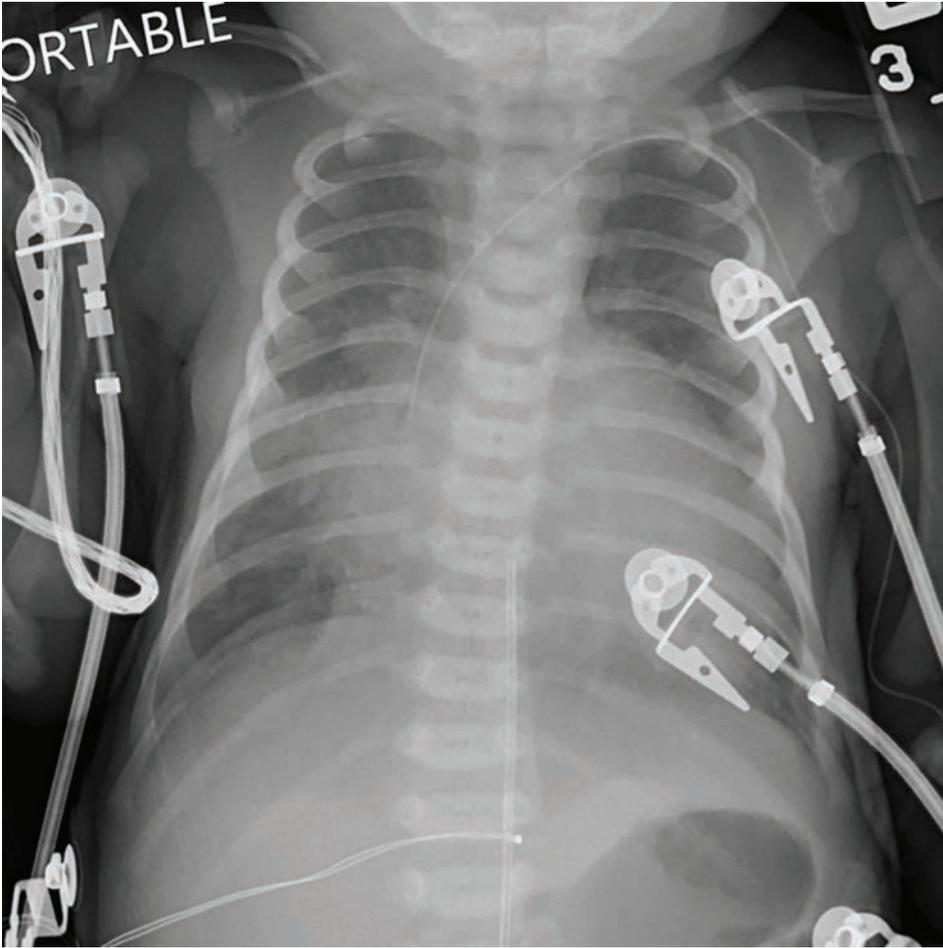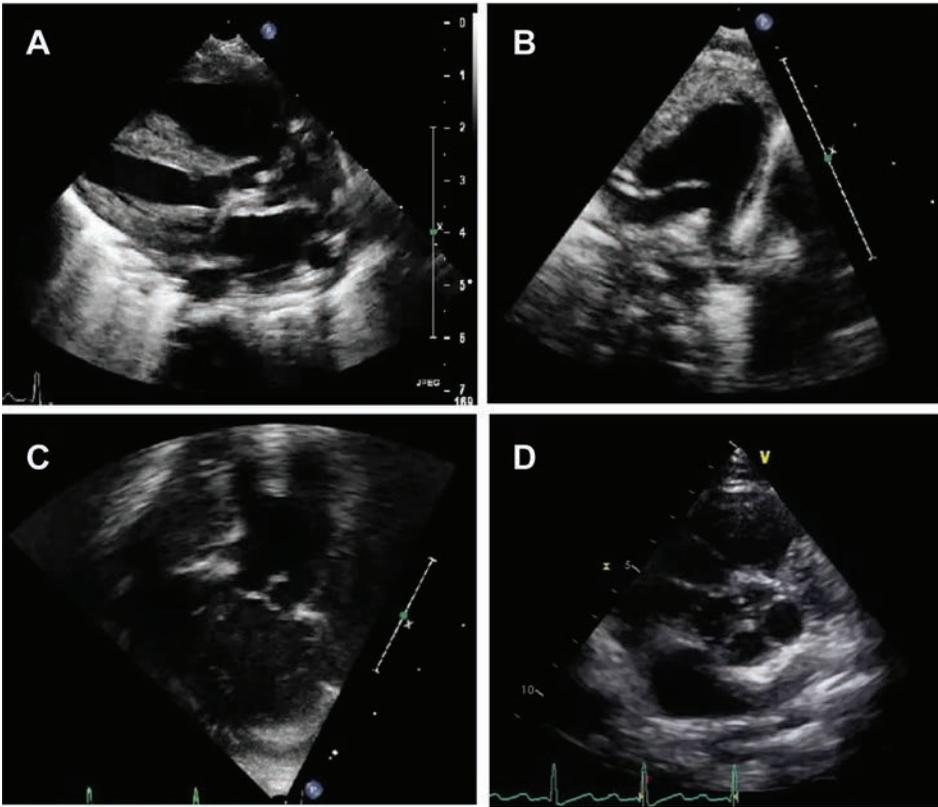
Figure 28-3. CXR of a patient with truncus arteriosus showing significant cardiomegaly, pulmonary congestion, and a narrow mediastinum due to the lack of thymus from DiGeorge syndrome.

disease. In addition, delaying surgical intervention does not provide any advantage for the repair. As such, patients with truncus arteriosus are repaired during the newborn period. Similarly, patients with AP window should be intervened upon at the time of diagnosis unless the window is small, in which case an elective repair can be offered.

Anesthetic Considerations

The chief consideration for the anesthetic management of these patients is to balance the pulmonary and systemic circulations. PVR is usually orders of magnitude lower than SVR, making these patients at best suffer from pulmonary overcirculation or at worst coronary ischemia. Invasive arterial pressure monitoring can help guide a slow,

Figure 28-4. Echocardiograms of patients with truncus arteriosus. A) Parasternal long-axis view showing a thickened and dysplastic truncal valve overriding a VSD. B) Suprasternal-notch view of a patient with truncus arteriosus type I demonstrating the origin of the MPA and branch PAs from the common trunk. C) Apical view demonstrating the common arterial trunk as well as the MPA segment arising from the ascending trunk. D) Parasternal-short axis view showing a quadricuspid truncal valve en-face. Images courtesy of Dr. Josh Kailin, www.pedecho.org.

controlled induction. Ventilation management should target keeping the PVR elevated: room air with gentle hypoventilation and hypercarbia. Subatmospheric oxygen (FiO$_2$ 18%) has been used historically but is not currently an accepted management strategy. ST-segment analysis and diastolic BP need to be closely monitored for any signs of ischemia. Vasoconstriction in the form of vasopressin can help keep diastolic BP in a comfortable range with the caveat that increasing SVR will decrease the systemic circulation. Once the sternum is open, the surgeon can snare the branch PAs to create a temporary "banded" circulation, improving systemic cardiac output and allowing greater latitude in increasing inspired oxygen. Assessment of the function, PVR, and SVR post-CPB can help guide decisions regarding what combination of pharmacologic therapies will best support the patient.

Surgical Repair

Truncus Arteriosus

The goal of the surgical repair of truncus arteriosus is to separate the PAs from the common trunk, repair the VSD by creating a baffle between the LV and the truncal valve, and establish continuity between the RV and the PAs with a valved conduit. The procedure is performed via median sternotomy with CPB, aorto-bicaval cannulation, and mild-moderate hypothermia. The PA branches are snared after initiation of CPB to avoid runoff into the lungs.

After cardioplegic arrest, the PA or branch PAs are separated from the common trunk. Depending on the anatomy, this may be achieved by transecting the common trunk and leaving a rim of aortic tissue around the PAs or harvesting the PAs as a button while visualizing the anatomy through an anterior aortic incision. It is important to recognize the relationship between the PAs and both the coronary arteries and the aortic valve in order to avoid injury, as they may be in close proximity. The truncal valve should be inspected and repaired if necessary. Truncal valve repair is challenging but there are different surgical techniques that can be used for this purpose. The coronary arteries are also inspected and may require intervention (e.g., unroofing in the case of intramural coronaries).

If the common trunk is transected, the aortic root and the distal ascending aorta are reanastomosed (an autologous pericardial patch may be necessary). If the PAs are harvested as a button, the defect is usually repaired with an autologous pericardial patch in order to avoid distortion of the coronary arteries and aortic valve.

A right ventriculotomy is performed and the VSD is closed toward the truncal valve with a piece of glutaraldehyde-treated autologous pericardium using either a series of interrupted pledgeted sutures or a continuous sutureline. A homograft RV-to-PA conduit is then placed. Due to the location of the ventriculotomy, the conduit tends to sit directly behind the sternum and may thus be smaller than the conduit used for other repairs.

Both an LA line and a peritoneal dialysis catheter are placed routinely. It is not customary to leave an open atrial communication in these patients.

In the case of a truncus arteriosus with an interrupted aortic arch, arterial cannulation is achieved using a graft sutured to the innominate artery (for the upper body) and a second cannula inserted into the PDA. The aortic arch is reconstructed using an aortic arch advancement technique (see Chapter 25). The rest of the procedure is performed as described for isolated truncus arteriosus.

AP Window

The surgical management of an AP window depends on the size and location of the window. In general, aortic cannulation is achieved on the distal ascending aorta or aortic arch. If the AP window is very distal, it may require placement of a graft on the innominate artery and performing the repair using antegrade cerebral perfusion (see Chapter 6). AP windows are usually repaired by dividing the window and closing each vessel with a patch of autologous pericardium to prevent distortion. However, each case should be individualized to the particular anatomic characteristics.

Postoperative Management

General Management

- **Fluids.** 25% maintenance with D5%/0.45% NS is standard. Careful attention should be paid to managing the patient with the barely necessary preload. Unnecessary preload may produce increases in myocardial wall stress and lead to further ventricular dysfunction and hypotension. In cases involving arch repair, the ischemic time may have been longer, thus potentially prolonging the period of possible postoperative LCOS.

- **Analgesia and sedation.** Analgesics and sedatives are adjusted for patient's comfort. For analgesia, a fentanyl infusion is used. Scheduled acetaminophen q6h (enteral, rectal, or IV) is used as an adjuvant. Sedation is achieved with a combination of dexmedetomidine and midazolam as a drip.

- **Vasoactive drugs.** Most patients will arrive from the OR on milrinone 0.25-0.75 mcg/kg/min, epinephrine 0.02-0.05 mcg/kg/min, and calcium chloride 5-15 mg/kg/hr (especially if suspected DiGeorge). Hypotension should be primary managed with inotropes when the LA pressure is higher than 10 mmHg.

- **Mechanical ventilation.** Patients are usually ventilated on SIMV-VC with pressure support, Vt 8-10 mL/kg and PEEP 5-7 cmH$_2$O, aiming for a pH of 7.35-7.45 and a SaO$_2$ >95%.

> **Truncus arteriosus repair (1995-2016)**
> Number of repairs: 99
> Median age at surgery: 17 days (4 days – 2.6 years)
> Median ICU length of stay: 10 days (3-74 days)
> Median hospital length of stay: 18 days (5-42 days)
> Perioperative mortality: 8%
> Mechanical circulatory support: none
>
> **Aortopulmonary window repairs (1996-2016)**
> Number of repairs: 26
> Median age at surgery: 22 days (6 days – 3 years)
> Median hospital length of stay: 13 days (4-60 days)
> Perioperative mortality: 4%
> Mechanical circulatory support: none

What to Expect in the First 24 Hours Postoperatively

The postoperative management is mainly directed to address possible LV and RV dysfunction from prolonged CPB time and pulmonary hypertension. If there is preoperative coronary insufficiency and there was coronary unroofing, there may be further systolic and diastolic dysfunction. Pulmonary hypertension should be managed with iNO and decreasing oxygen consumption with sedation and intermittent muscle relaxation.

- **Ventilation.** Transitioning from the OR, the lungs will be significantly improved from the preoperative period secondary to continuous ultrafiltration. There may be mild pulmonary hemorrhage that should managed with PEEP 5-10 cmH$_2$O.

- **Fluids.** Even to slightly negative. Peritoneal dialysis is instituted on arrival to the CICU. If additional fluids/colloids are needed, one should carefully titrate small-volume boluses (1-5 mL/kg/dose), not to exceed LAP >12 mmHg.

- **Nutrition.** Should write for TPN the day after surgery. If DiGeorge syndrome is suspected or confirmed, there may be oropharyngeal dysfunction and OT should be consulted before starting feeds (see Chapter 58).

Complications

- **LCOS.** Primarily treated with inotropes. A combination of low-dose epinephrine and standard-dose milrinone.
- **Pulmonary hypertension.** It should be managed initially with iNO and potentially other advanced therapies (see Chapter 79).
- **Chylothorax.** May be secondary to a combination of pulmonary hypertension, RV dysfunction, and PA branch stenosis. Also, patients with DiGeorge syndrome have a higher incidence of chylothorax. For management of chylothorax see Chapter 77.
- **Mechanical circulatory support.** The use of mechanical support is rare after truncus arteriosus repair, except when there is significant preoperative ischemia with severe ventricular dysfunction.
- **PA branch stenosis.** Due to the size of the truncal root, the right PA may be compressed behind the reconstructed truncal root. This possible complication should be monitored using echocardiography.

Long-Term Follow-Up

Close follow-up of patients with truncus arteriosus and routine echocardiographic surveillance is mandatory. Long-term prognosis will be partly determined by the status of the truncal valve. Patients with truncal valve insufficiency, in particular those that underwent truncal valve repair during the initial operation, are at high risk of requiring truncal valve replacement in the future. Most patients will require RV-PA conduit replacement in the first 3-5 years of life. Larger conduits placed at future operations tend to have a longer longevity.