Chapter

67 Truncus Arteriosus

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A four-day-old term baby boy is transferred from an outside hospital with a postnatal observation of a murmur. There was no prenatal care prior to delivery. The baby had some peripheral cyanosis at birth which corrected with supplemental oxygen. His tachypnea is more pronounced with feeds.

Vitals BP 63/39, Pulse 166, Temp 36.9°C, Resp 49/min at rest and 60–70 per min with feeds, Wt 2.8 kg, SpO_2 94% on 1L nasal cannula.

Physical examination reveals a cleft palate, coarse respiratory sounds, and a continuous precordial murmur. Subcostal retractions are present when the child is fed. The abdomen is soft with a fullness appreciated over the hepatic border. Bounding pulses are present throughout.

What Is a Truncus Arteriosus (TA) Anomaly?

Truncus arteriosus (TA) is a rare congenital anomaly characterized by a common arterial trunk from which the ascending aorta and the pulmonary arteries originate distal to the coronary arteries but proximal to the first brachiocephalic branch of the aortic arch (Figure 67.1).

The common arterial trunk arises from normal ventricles and the semilunar valve is labeled as a truncal valve. In patients with a normal aortic arch, the ductus arteriosus may be either absent or diminutive.

How Is TA Classified?

The pulmonary artery arises from the TA in many different patterns and is the basis used to classify subtypes of TA. There are two classifications commonly used: Collett and Edwards (1949) and Von Praagh (1965).

The earliest classification, developed by Collett and Edwards, includes TA types I–IV, as follows:

- Type I TA is characterized by origin of a single pulmonary trunk from the left lateral aspect of the common arterial trunk, with the left and right pulmonary branches arising from the pulmonary trunk.
- Type II TA is characterized by separate but proximate origins of the left and right pulmonary arterial branches from the posterolateral aspect of the common arterial trunk.
- Type III TA occurs when the branch pulmonary arteries originate independently from the common arterial trunk or aortic arch, most often from the left and right lateral aspects of the trunk.
- Type IV TA, originally proposed as a form of the lesion with neither pulmonary arterial branch arising from the common trunk, is now recognized to be a form of pulmonary atresia with ventricular septal defect.

The Van Praagh classification includes four primary types of TA variants:

- Type A1 is identical to the type I of Collett and Edwards.
- Type A2 includes Collett and Edwards type II and most cases of type III, namely those with separate origin of the branch pulmonary arteries from the left and right lateral aspects of the common trunk.
- Type A3 includes cases with origin of one branch pulmonary artery (usually the right) from the common trunk, with pulmonary blood supply to the other lung provided either by a pulmonary artery arising from the aortic arch (a subtype of Collett and Edwards type III) or by hyphenate arterial collaterals.
- Type A4 is defined not by the pattern of origin of branch pulmonary arteries, but rather by the coexistence of an interrupted aortic arch. In the

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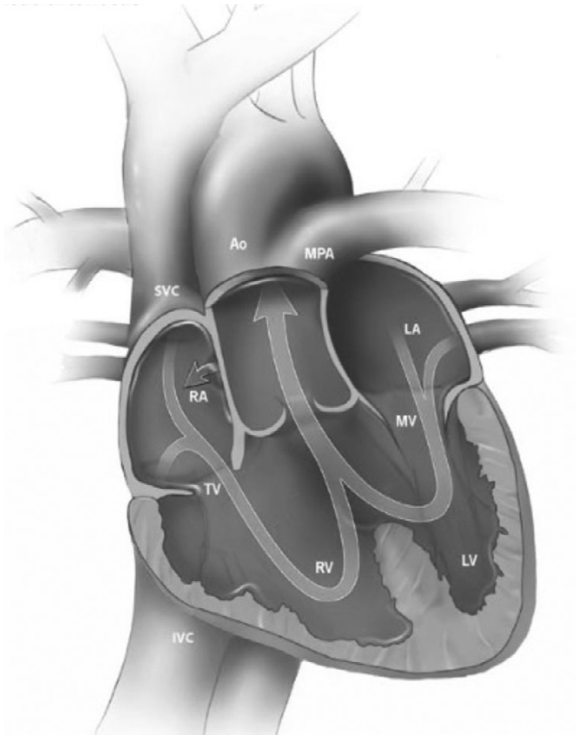

Figure 67.1 Illustration of truncus arteriosus (TA). RA and LA, right and left atrium; RV and LV, right and left ventricles; SVC and IVC, superior and inferior vena cava respectively; TV, MV, tricuspid, mitral, valves respectively; MPA, main pulmonary artery; Ao, aorta. Image courtesy of Centers for Disease Control and Prevention, USA

vast majority of cases of type A4, which fall into the type I of Collett and Edwards, the pulmonary arteries arise as a single pulmonary trunk that then branches. In any of these patterns, intrinsic stenosis, hypoplasia, or both may be present in one or both branch pulmonary arteries, which may have an effect on management and outcome.

What Are the Presenting Signs and Symptoms?

Poor feeding, diaphoresis, tachypnea, and cyanosis are common presenting symptoms with TA although many are prenatally diagnosed by echocardiography.

What Is the Embryological Basis for This Anomaly?

The failure of complete or incomplete septation of the embryonic TA leads to this uncommon anomaly. Because the common trunk originates from both the left and right ventricles, and pulmonary arteries arise directly from the common trunk, a ductus arteriosus is not required to support the fetal circulation.

Is There a Genetic Abnormality Associated with TA?

As with other congenital cardiac anomalies of the conotruncal region, a substantial number of patients with TA (approximately 30–40%) have microdeletions within chromosome band 22q11.2. This particular type of chromosomal deletion is thought to affect migration or development of cardiac neural crest cells and may contribute to the pathogenesis of TA.

Is TA Associated with Any Other Congenital Heart Defects?

Structural abnormalities of the truncal valve, including dysplastic and supernumerary leaflets including truncal valve regurgitation (moderate or severe) may be present in 20% or more of patients. Approximately one third of patients will have a right-sided aortic arch.

Coronary artery abnormalities are common and may include as single coronary artery with an intramural course as the most important variation.

Interrupted aortic arch is a commonly noted anomaly, which almost always occurs between the left common carotid and subclavian arteries (type B).

Other uncommon associations include: persistent left superior vena cava, aberrant subclavian artery, and atrial septal defects. Complete atrioventricular septal defect, double aortic arch, and various forms of functionally univentricular heart are some rare associations.

What Is the Typical Pathophysiology of the Anomaly?

Cyanosis and systemic ventricular volume overload are the two hallmarks of TA.

The cardiac output from both ventricles is directed into the common arterial trunk – arterial trunk. The amount of pulmonary blood flow derived from this combined ventricular output is dependent on the ratio of resistances to flow in the pulmonary and systemic vascular beds. The mixing of left and right ventricular output at the level of the arterial trunk (during systole) is the reason for cyanosis.

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Infants with TA may present in shock because of high output heart failure with significant pulmonary overcirculation. This may resemble the presentation of neonatal sepsis, especially when the ratio of Qp:Qs is sufficiently high that the patient is not cyanotic.

What Is the Natural History of TA Anomaly?

In numerous earlier series, the median age at death without surgery ranged from two weeks to three months, with almost 100% mortality by age one year. Cardiac arrest or multiple organ failure secondary to systemic hypoperfusion, progressive metabolic acidosis, and myocardial dysfunction are causes of death in the unrepaired patient with TA.

Currently, for patients undergoing complete repair in the neonatal period or early infancy, early postoperative mortality is generally less than 10%. Among patients surviving the initial postoperative period, the survival rate at a 10- to 20-year follow-up is higher than 80%, with most deaths resulting from sequelae of late repair (pulmonary vascular obstructive disease), reinterventions, or residual/recurrent physiologic abnormalities.

Is This a Surgical Emergency? What Is the Surgical Approach?

The degree of pulmonary overcirculation leading to congestive heart failure dictates the urgency and need for medical management versus surgical repair.

Early neonatal repair is the recommendation. However, a delay of up to two to three months is based on the individual. Early repairs avoid the development of severe pulmonary vascular occlusive (from pulmonary overcirculation) disease and the increase in mortality.

The repair involves removal of the pulmonary arteries, and closure of the defect either directly or with a patch, closure of the VSD, and a conduit from the RV to the pulmonary arteries to supply pulmonary blood flow.

What Are the Preanesthetic Concerns in This Child with TA Scheduled for Surgery?

DiGeorge syndrome, now referred to as 22q11 deletion syndrome and velocardiofacial syndrome (Shprintzen syndrome) is associated in 30–35% of patients with TA. The patients have variable, microdeletions of chromosome 22q11 or CATCH-22 syndrome.

The CATCH-22 acronym denotes the common associated anomalies:

C – Cardiac anomalies – especially interrupted aortic arch, TA, and tetralogy of Fallot

A – Abnormal facies (velocardiofacial syndrome)

T – Thymic aplasia

C – Cleft palate

H – Hypocalcemia/hypoparathyroidism

22 – Deletion of the 22nd chromosome

Patients with 22q deletion syndrome (previously DiGeorge syndrome) have thymic aplasia and parathyroid dysfunction and are at risk for developing hypocalcemia and immune deficiencies. They also have characteristic facies consisting of a cleft palate, long narrow face, prominent maxilla and a retruded chin. A careful airway assessment is necessary as these patients are often difficult intubations.

Other noncardiac anomalies found sporadically in patients with TA include renal abnormalities, vertebral and rib anomalies, and anomalies of the alimentary tract.

The association of the above syndromes requires vigilant monitoring of the serum calcium levels to avoid the risk of hypocalcemia. The possibility of immune deficiencies requires blood products to be irradiated.

Is There Any Role for Observing Infection Precautions in These Patients?

Thymic absence poses a risk for immune deficiencies in the form of T cell mediated immunity. This requires that all blood products be irradiated and serum calcium levels monitored when citrated blood products are administered. Sterile techniques should be employed during the placement and conduct of procedures.

How Is Anesthesia Managed in These Critically Ill Patients?

The patient's age and anatomy of the lesion dictate the anesthetic management. Patients with severe congestive heart failure (CHF) may require preoperative

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inotropic support, and anesthetic induction in these patients is aimed at maintaining systemic vascular resistance (SVR) and preserving myocardial function.

Some of these patients may be intubated in the ICU because of cardiopulmonary compromise.

The goal is to balance pulmonary to systemic blood flow by manipulating factors that enhance or reduce pulmonary blood flow. This becomes more critical after the first few days of life when a natural reduction in pulmonary vascular resistance (PVR) occurs promoting excessive pulmonary blood flood.

These infants are ventilated with an FiO_2 of 21% and $PaCO_2$ maintained around 45–50 mmHg targeting an arterial pH of 7.25–7.35. Transporting these patients to the operating room requires that the FiO_2 is maintained around 21%, with hypoventilation to achieve hypercarbia. Analgesia and muscle relaxants should be chosen to avoid tachycardia and hypotension. Inotropic support should be continued during transport to the operating room in infants with CHF.

The non-intubated infant must be managed very carefully. Intravenous induction is preferred with a synthetic opioid and muscle relaxant. Mask ventilation with 100% oxygen may be used immediately prior to intubation, however, once the trachea is intubated the FiO_2 should be decreased to 0.21 targeting a saturation of 75–85%. Minute ventilation should be controlled to maintain a $PaCO_2$ of 45–50 mmHg.

Maintenance of anesthesia and ventilation should be carefully titrated to optimize a balance between PVR and SVR, and a Qp:Qs ratio of 1:1 (see Chapter 62). Hyperventilation and high inspired oxygen concentrations should be avoided as they promote a decrease in PVR and will exacerbate pulmonary blood flow and lower diastolic perfusion pressure. In patients with truncal valve regurgitation this may precipitate myocardial ischemia.

Some patients may present in late infancy and may have developed significant pulmonary hypertension from chronic pulmonary overcirculation. Ironically these patients will require higher concentrations of FiO_2 to maintain SaO_2 between 75–85%.

These patients will require inotropic support and afterload reduction upon weaning from cardiopulmonary bypass. Transesophageal echocardiography aids in assessment of ventricular volumes and pulmonary artery pressures with the intent to improve right heart function. Pulmonary hypertension is frequently noted after weaning from cardiopulmonary bypass and may continue into the postoperative period.

What Are the Postoperative Concerns Following Repair of TA?

These patients should be sedated, paralyzed, and ventilated in the intensive care unit during the first 24 hours to avoid and/or minimize pulmonary hypertensive crises. Right heart inotropic support (milrinone), hyperventilation, 100% oxygen FiO_2, correction of acidosis, and nitric oxide are used as needed to treat any pulmonary hypertensive crisis. Residual VSDs, truncal valve anomalies (stenosis/regurgitation), or right ventricular incision (VSD closure, RV-PA conduit) may also cause right heart dysfunction. A bedside transthoracic echocardiography may be helpful in resolving this treatable clinical dilemma. Postoperative arrhythmias such as right bundle branch block, complete heart block, and atrial or junctional ectopic tachycardia can cause ventricular dysfunction. Supplemental calcium infusions may be beneficial due to the common association of TA with DiGeorge syndrome and after administration of citrate anticoagulated blood products.

What Are the Sequelae of TA Repair?

Residual VSD, truncal valve regurgitation, and stenosis of RV-PA conduits are late sequelae of truncus repair.

Suggested Reading

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