

What is Truncus Arteriosus?

Truncus arteriosus is a rare, complex form of congenital heart disease (CHD), accounting for about 1 percent of CHD. It is slightly more common in males. It is a cyanotic lesion. Mixing of red (oxygenated) and blue (unoxygenated) blood cause cyanosis or blueness.

There are two great vessels in a normal heart. The pulmonary artery takes blood from the right ventricle to the lungs. The aorta pumps blood from the left ventricle to the body. In truncus arteriosus, there is only one great vessel. It gives rise to the aorta, the pulmonary artery and the coronary arteries. The defect always sits over a large ventricular septal defect (VSD).

There are some adults who are undiagnosed and/or have unrepaired truncus arteriosus. They do not do as well as those who are repaired. They are at high risk for pulmonary hypertension and Eisenmenger syndrome.

How does Truncus Arteriosus develop?

The heart starts out as a tube. As the tube rotates, it gives rise to the different parts of the heart. This occurs in the first 5-6 weeks of development. The pulmonary artery and aorta come out of the head of the tube. Truncus starts as one big blood vessel. During development the wall between the pulmonary artery and aorta does not form. You end up with one big vessel or trunk.

There are four different types of truncus.

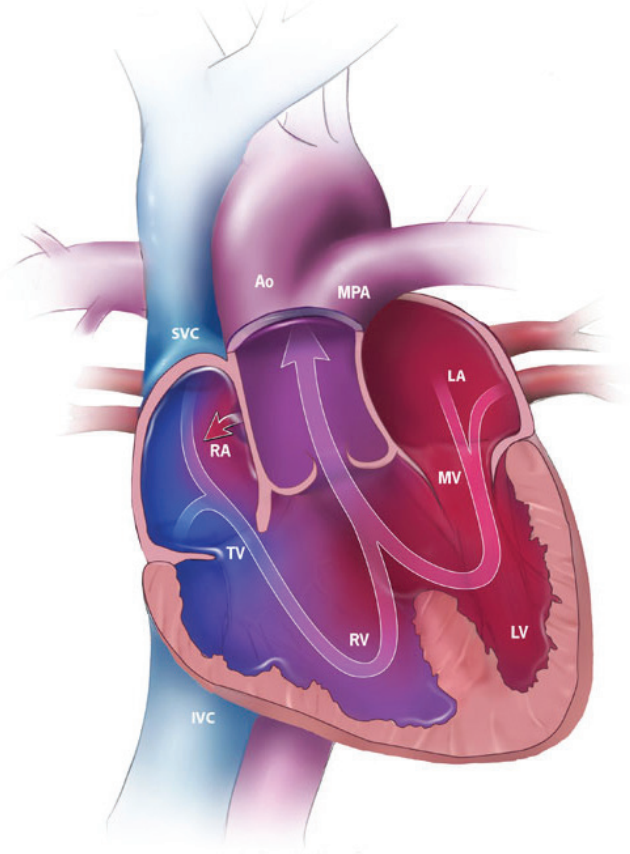
- In type I, there is a single pulmonary trunk, known as a nubbin. This comes off the left side of the common trunk. The left and right branches of the pulmonary artery arise from that nubbin.
- In type II, the left and right pulmonary artery branches have separate sites of origin. Both come off of the rear side of the common trunk.
- In type III, the left and right branches of the pulmonary arteries come off either the left or right side of the common trunk independently.
- Type IV is the most complex form. It is now recognized as a form of pulmonary atresia. This is a condition in which the pulmonary valve fails to form properly. In this type, the pulmonary arteries come off of the descending portion of the aorta.

Truncus Arteriosus can be associated with other lesions. Among these are interruption of the aorta, VSD and DiGeorge syndrome (22q11).

How does Truncus Arteriosus present?

Truncus Arteriosus is almost always diagnosed in infancy. Infants present with poor feeding, sweating, poor weight gain, breathing problems, cyanosis and/or congestive heart failure. These children have surgery to correct the defect at an early age.

Truncus Arteriosus



RA – Right Atrium	IVC – Inferior Vena Cava
RV – Right Ventricle	MPA – Main Pulmonary Artery
PA – Pulmonary Artery	Ao – Aorta
LA – Left Atrium	TV – Tricuspid Valve
LV – Left Ventricle	MV – Mitral Valve
SVC – Superior Vena Cava	

Courtesy of the Centers for Disease Control and Prevention, National Center on Birth Defects and Developmental Disabilities

There are some adults who are undiagnosed and/or have unrepaired truncus arteriosus. They do not do as well as those who are repaired. They are at high risk for pulmonary hypertension and Eisenmenger syndrome. This is the result of years of increased pressure on the heart.

How is Truncus Arteriosus repaired?

Truncus Arteriosus surgery usually occurs in the first few weeks of life. The surgery involves separating the pulmonary arteries from the main trunk. The truncus then becomes the first part of the aorta. The VSD is closed. A connection is made between the right ventricle and pulmonary arteries. This is done by using a conduit. As the child grows, the conduit will need to be replaced.

Truncus Arteriosus is a highly complex heart defect. This means that experts recommend that you receive care from an ACHD specialist throughout your life, even if you are feeling fine.

What are the long term outcomes for adults with Truncus Arteriosus?

While there are some adults with unrepaired truncus arteriosus, many of the problems that adults face are the result of complications of the repair itself and the underlying CHD.

While uncorrected patients do not do well, patients who have had a primary repair generally do very well.

Problems that develop in adulthood may result from:

1. The age of the initial repair
2. Number of surgeries
3. Symptoms
4. Size of the VSD
5. Size of the pulmonary arteries
6. If the truncal valve is narrow or leaking
7. If there is pulmonary hypertension
8. If there are other associated heart problems

Most issues that adults develop are the result of complications from their primary surgery. These include:

1. Truncal stenosis or regurgitation
2. Stenosis or regurgitation of the right ventricle to pulmonary artery conduit(s)
3. Ventricular dysfunction
4. Arrhythmias or irregular heartbeats
5. Pulmonary hypertension or high blood pressure in the lungs, if untreated or if there was late repair

What tests are usually done?

Your doctor will most likely give you an Electrocardiogram (ECG). This test can tell your doctor if you have arrhythmias.

He/she might ask you to wear a Holter or event monitor. These are also helpful in detecting atrial or ventricular arrhythmias over a period of time.

Imaging is very important. Your doctor will probably want you to have an echocardiogram. This will show what the heart looks like after surgery. It can tell your doctor if you have truncal narrowing or stenosis.

As an adult, your doctor might order a cardiac CT and/or MRI to look at the size and function of your ventricles. These tests can also tell your doctor if you have leaking or narrowing in the RV-PA conduit or neo-aortic valve.

What are my treatment options as an adult?

Your treatment as an adult will depend on the specific nature and severity of the problem. If your conduit becomes narrow or leaky, it may be necessary to replace or repair it. This is done in one of two ways:

1. Your doctor may decide to put in a Melody valve. This is an artificial heart valve that is inserted using a cardiac catheter
2. Surgical valve replacement may be necessary if the conduit is damaged or cannot be reached with the catheter.

Either way, before doing surgery your doctor needs to know if you have pulmonary hypertension.

Arrhythmias are managed as they would be in any adult CHD patient. Your doctor will figure out the cause of the arrhythmia. He/she may then put you on a medication like a beta blocker and/or anti arrhythmia medicine. In some cases, an ablation may be needed or you may need an Implantable Cardioverter Defibrillator (ICD). If you have pulmonary hypertension, medications are also available to treat this.

Can I have a baby?

Pregnancy is possible in some patients with repaired truncus arteriosus. It is very important that you see an adult congenital heart disease (ACHD) specialist if you are thinking about having a baby and before you get pregnant. A successful pregnancy requires a cardiac specialist with expertise in this area and a high risk obstetrical physician. If you do become pregnant, your ACHD cardiologist and high risk obstetrician will take special steps to protect your heart during your pregnancy and delivery.

What type of cardiac care is recommended for adults with Truncus Arteriosus?

According to the American Heart Association and the American College of Cardiology, Truncus Arteriosus is a highly complex heart defect. This means that experts recommend that you receive care from an ACHD specialist throughout your life, even if you are feeling fine.