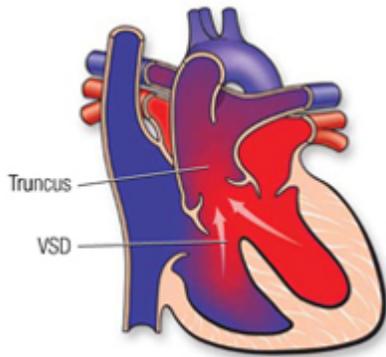


Truncus Arteriosus



What is it?

Truncus arteriosus is characterized by a large ventricular septal defect over which a large, single great vessel (truncus) arises. It occurs when the two large arteries carrying blood away from the heart don't form properly and one large artery is present instead. This single great vessel carries blood both to the body and to the lungs. This artery (the truncus) sits over a large opening or hole in the wall between the two pumping chambers (ventricular septal defect).

What causes it?

Some patients with truncus arteriosus have a condition called DiGeorge syndrome, however a known genetic cause is only found in a minority of patients.

How does it affect the heart?

The blood leaving the heart can go the heart or lungs. Early in life this results in quite a bit of extra blood going to the lungs, which makes the heart work very hard. Over time the extra blood flow damages the blood vessels in the lungs, resulting in pulmonary hypertension. When this happens (often before one year of life) the patient becomes blue, and it's often too late to do successful surgery. The valve leading to the main blood vessel can have blockage or leakage, which can also make the heart work harder.

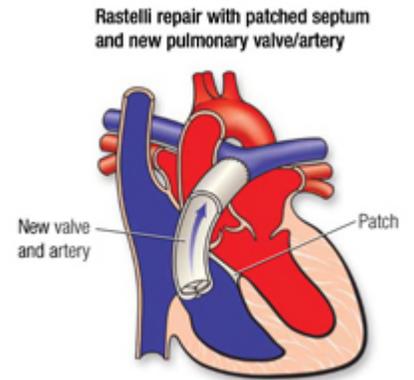
How does truncus arteriosus affect me?

Truncus arteriosus causes symptoms of heart failure early in life. If it's not repaired, severe pulmonary hypertension occurs. If, like almost all adults with this condition, you've had surgery, the most important issues are related to the artificial connection between the heart and lungs (pulmonary valve and conduit) and whether the truncal valve (the valve between the left ventricle and the body after surgery) has a significant amount of leakage. If the pulmonary valve has significant leakage or blockage, symptoms could develop (including exercise intolerance and heart rhythm issues).

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If my defect was repaired in childhood, what can I expect?

Almost all adults with truncus arteriosus had surgery in childhood. Surgery is required to close the ventricular septal defect and separate blood flow to the body from blood flow to the lungs. This is generally done early in infancy to prevent high blood pressure from damaging the lungs' arteries. A patch is used to close the ventricular defect. The pulmonary arteries are then disconnected from the single great vessel and a tube (a conduit with a valve) is placed from the right ventricle to these pulmonary arteries. This is sometimes called a Rastelli repair.



The conduit and valve will need to be replaced at least two or three times in childhood and may also need to be replaced in adulthood. There are new catheter-based procedures that may allow the pulmonary valve to be replaced without surgery. In a smaller number of patients the truncal valve may also need to be repaired or replaced surgically.

Problems you may have

Heart Function

In the long-term period after the operation, the heart muscle's ability to contract may decrease. You may need medication including diuretics, agents to help your heart pump better and drugs to control your blood pressure.

Heart Rhythm Disturbances

Patients with repaired truncus arteriosus have a higher risk for heart rhythm disturbances called arrhythmias. These can originate from the atria (the heart's two upper chambers) or the ventricles (the two lower chambers). Sometimes they may cause dizziness or fainting. Medication may be required to control them. In rare cases, a procedure in the cardiac catheterization laboratory or the operating room may be required to eliminate these arrhythmias and control symptoms.

High Blood Pressure in the Lungs (Pulmonary Hypertension)

Sometimes, even when the defect is repaired early, pulmonary hypertension becomes progressively worse. Patients may experience shortness of breath, decreased exercise endurance and sometimes headaches and dizziness.

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Ongoing Care:

What will I need in the future?

Patients with truncus arteriosus need regular follow-up with a cardiologist with special training in adult congenital heart disease. You may need to take medicine after your operation to help your heart pump better. You should also consult a cardiologist with expertise in caring for adults with congenital heart disease if you are undergoing any type of non-heart surgery or invasive procedure.

Medical Follow-up

Your cardiologist will monitor you with a variety of tests. These include electrocardiograms, Holter monitors, exercise stress tests and echocardiograms to determine when another procedure such as cardiac catheterization may be needed.

Activity Restrictions

If you have decreased heart function or rhythm disturbances, you may need to limit your activity. If valve obstruction and leakage is mild and tests show good heart function and no abnormal heart rhythms, you can usually participate in some sports. Your cardiologist may recommend avoiding certain intense competitive sports. Ask your cardiologist which activities are appropriate.

Endocarditis Prevention

People who have truncus arteriosus require endocarditis prophylaxis.

Pregnancy

Women with repaired truncus arteriosus may handle pregnancy well. However, some who have significant problems with their pulmonary or truncal valve or have significant pulmonary hypertension may be at much higher risk. These women should be followed in centers with high risk obstetrical services and adult congenital heart disease expertise.

Will I Need More Surgery?

Conduit Replacement

The conduit connecting the right ventricle to the pulmonary artery conduit may become obstructed (stenotic) over time and may need to be replaced (see above). Timing of the replacement varies. The peripheral pulmonary arteries also may become narrowed and require treatment.

Sometimes conduits and peripheral pulmonary artery obstructions may be dilated using a balloon-tipped catheter or an expandable stent in the cardiac catheterization laboratory. This procedure may help extend the time between conduit changes. Sometimes surgery is required to enlarge the narrowed area. Your cardiologist will decide whether a balloon/stent procedure or surgery is best for you.

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Truncal/Aortic Valve Replacement

The functional aortic valve is actually the large truncal valve from the single vessel, which arose over the ventricular septal defect before surgical repair. This valve sometimes becomes leaky over time and may need to be replaced.