

What is Truncus Arteriosus?

Truncus Arteriosus is a very rare condition of the heart that is very serious and lethal if not repaired. It is caused very early in the pregnancy by the failure of the common truncus (which forms the arteries of the heart) to separate into the aorta and pulmonary artery. Normally, the aorta is the vessel coming out of the heart that is attached to the left ventricle and carries blood with oxygen to the head, arms, legs, and organs of the body.

The pulmonary artery is attached to the right ventricle and carries blood without oxygen to the lungs (where it then picks up oxygen). Truncus Arteriosus occurs in less than one in ten thousand live births.

Truncus Arteriosus is usually picked up early in life because the baby will look blue to the nurse or the doctor shortly after it is born. Also shortly after birth it is common for someone to hear a murmur (which is an extra sound you hear when you listen to the baby's heart). This murmur is there because almost always there is a large hole in the center of the heart or because the valve of the truncus (normally the aortic valve) is not properly formed. The pulmonary arteries that supply blood to the lungs come off of the Truncus rather than from the right ventricle. Because of all the mixing of blood within the heart that occurs with this condition, the heart has to work extra hard and rapidly goes into heart failure (this means the heart cannot pump all the blood it needs to, not that it stops!) within weeks. This causes the baby to breathe rapidly and not eat well, and eventually to not gain weight. Surprisingly, sometimes these babies can look pink. We oftentimes have to give these babies medicines like Digoxin and Lasix to help the heart beat more efficiently.

Truncus Arteriosus must be surgically fixed early in life in most cases, usually within the first three weeks of life.

If it is done early there is a high likelihood that there will be damage to the baby's lungs because of the extra amount of blood flow through them. Very rarely there is enough narrowing of the pulmonary arteries that surgery can be postponed. Very frequently there is an abnormal aortic valve present that either is narrowed or leaks. If the valve is leaking too much it needs to be replaced at the time of surgery. Thus, it is possible to require both aortic and pulmonic valves to be replaced.

Because surgery is required at such a young age it is usually necessary to have additional surgery at 3 to 7 years of age as the original valves do not grow. Patients with Truncus require a lifetime of follow-up because of the need to periodically replace valves and distortion of the pulmonary arteries that may cause problems as they get older.

Truncus Arteriosus is frequently associated with a condition called DiGeorge Syndrome which can cause problems with the immune system. Also there are chromosomal abnormalities associated with deletions of chromosome 22 seen with Truncus Arteriosus. This defect can be seen in girls who become pregnant who use Accutane for the treatment of acne.

If you have any questions, please ask one of