Truncus Arteriosus

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

Some children with truncus arteriosus have a condition called DiGeorge Syndrome.

Can it be repaired?
Surgery is necessary to close the ventricular septal defect and separate blood flow to the body from blood flow to the lungs. This is usually done early in infancy to prevent high blood pressure from damaging the lung arteries. A patch is used to close the ventricular defect. The pulmonary arteries are then disconnected from the single great vessel (the truncus) and a tube (a conduit or tunnel) is placed from the right ventricle to the pulmonary arteries. This is sometimes called a Rastelli repair.

Will my child need more surgery?
The conduit connecting the right ventricle to the pulmonary artery conduit may become narrowed and blocked (stenotic) over time, or the child may outgrow the conduit. It may have to be replaced from time to time. Timing of the replacement varies. The peripheral pulmonary arteries also may become narrowed and require treatment. Sometimes conduits and peripheral pulmonary artery narrowings 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 operations for conduit changes. Sometimes surgery is required to enlarge the narrowed area. Your child’s cardiologist will discuss whether a balloon/stent procedure or surgery is best.
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The 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.

What ongoing care will my child need?
Children with truncus arteriosus need regular follow-up with a pediatric cardiologist and they may need to take medicine after surgery. Your child’s cardiologist will evaluate with a variety of tests including electrocardiograms and echocardiograms to determine when another procedure such as cardiac catheterization may be needed.

What activities will my child be able to do?
If valve obstruction and leakage is mild and tests show good heart function and no abnormal heart rhythms, your child can usually participate in some sports. Your cardiologist may recommend avoiding certain intense competitive sports. Ask your child’s cardiologist which activities are appropriate.

What about preventing endocarditis?
For patients with uncorrected or partially corrected truncus arteriosus, antibiotics are recommended before certain dental procedures to prevent endocarditis. S