Transposition of the Great Arteries, D-Type

What Is It?

In this defect, which accounts for 5% of the cases of congenital heart disease, the two main arteries (aorta and pulmonary artery) are connected to the wrong chambers of the heart. This condition is more common among males, affecting 3 boys to every 1 girl.

In TGA, the aorta leaves the right ventricle (rather than the left as in a normal heart) and takes blue (unoxyninated) blood to the body while the pulmonary artery leaves the left (rather than right) ventricle and takes red (oxygenated) blood to the lungs. This defect will require surgery, usually in the first week of life.

What Are Its Effects?

Transposition of the Great Arteries, D-Type
1) Patent (open) Foramen Ovale
2) Transposition of the Pulmonary Artery and Aorta
3) Patent (open) Ductus Arteriosus

Normal Heart
After birth, the infant is kept alive by the mixing of oxygenated blood from the left atrium with unoxygenated blood in the right atrium. This is possible because of an opening called the Foramen Ovale (1 in the diagram at left below) in the atrial septum (the muscle wall that divides the two atria). The Foramen Ovale provides one way for blood to mix, but it is often not enough. The Ductus Arteriosus (2), which connects the aorta and pulmonary artery in the fetal and newborn heart, also provides a way for deoxygenated (blue blood) to mix with the oxygenated (red blood).

The Foramen Ovale and Ductus Arteriosus are features of the fetal heart that usually close soon after birth. Though these may allow enough mixing of blood to keep an infant alive initially, Transposition of the Great Arteries would still prove fatal if measures are not taken to increase the amount of this mixing of the two circulations.

Transposition of the Great Arteries, D-Type

1) Patent (open) Foramen Ovale
2) Patent (open) Ductus Arteriosus
How Is It Treated?

At birth, the baby with TGA is usually stable. However careful examination often reveals that there is some degree of cyanosis (blueness). If the ductus arteriosis begins to close or if the Foramen Ovale becomes restrictive, then the infant can show signs of worsening cyanosis and respiratory distress. Drugs (such as prostaglandin E1) may make the infant less blue by causing the ductus arteriosus, a small blood vessel that connects the aorta with the pulmonary artery, to reopen.

If oxygen levels are still too low after prostaglandin therapy, or if the infant is very ill, temporary relief may be achieved by performing a procedure called a Balloon Septostomy (see illustrations below) in the catheterization lab.

The Balloon Septostomy involves the insertion of a tube (catheter) with an uninflated balloon at its end from a leg vein into the heart and through the foramen ovale into the left atrium. The balloon is then inflated and withdrawn, tearing the atrial septum as it is pulled back into the right atrium. This large opening allows for better mixing of the unoxygenated (blue) and oxygenated (red) blood.
A Balloon Septostomy in the catheterization lab is only a temporary measure, however. When the infant is about one week old, an Arterial Switch Operation is performed which corrects the defect by connecting the aorta to the left ventricle and the pulmonary artery to the right ventricle, as in a normal heart (see illustration below).

This procedure also involves reconnecting the small coronary arteries so that the heart muscle receives red (oxygenated) blood. The foramen ovale is also closed at this time and the patent ductus arteriosus (PDA) is divided and closed off. (yellow circles)
Surgical Repair of Transposition of the Great Arteries, D-Type (Jatene Arterial Switch)
The positions of the great arteries are switched and the foramen ovale and patent ductus arteriosus (yellow circles) are closed off.
Circulation with Transposition of the Jatene Arterial Switch
Oxygen-rich blood is shown in red, oxygen-poor blood in blue