

Transposition of Great Vessels

Transposition of the great vessels also called dextro-transposition of great vessels, is a cyanotic congenital heart lesion that is characterized by the aorta arising from the right ventricle and the pulmonary artery arising from the left ventricle. This defect creates two separate circulatory systems which run in parallel, resulting in deficient oxygen supply to the body and severe cyanosis. Diagnosis is typically made via echocardiography. Initial treatment consists of Prostaglandin E1 (PGE1) in order to keep the ductus arteriosus open (PDA) until surgical intervention can reverse the malpositioned vessels. Other treatment includes balloon atrial septostomy, arterial switch operation, and atrial switch operation.



PLAY PICMONIC

Pathophysiology

Pulmonary and Systemic Circulation are in Parallel

Circulation of Lungs and Body are separate and Parallel

Transposition of the great vessels occurs when the aorta arises from the right ventricle and pulmonary artery arises from the left ventricle, due to failure of the aorticopulmonary septum to spiral. Deoxygenated blood from the body returns to the right atrium, but is then pumped back to the body via the aorta, without ever picking up oxygen in the lungs. Oxygenated blood from the lungs is continuously pumped back into the lungs by the pulmonary artery, without ever going to the body. This creates two separate circulatory systems, which are classically described as running in parallel, instead of the normal circulatory system, which runs in series.

Incompatible with Life Without a Shunt

Lifeguard can't save a Life without Shunting blood

This circulation is incompatible with life unless there is communication and mixing between the two parallel circuits. Mixing between the two circuits can occur via extracardiac connections such as a patent ductus arteriosus (PDA) or via intracardiac connections such as a patent foramen ovale, atrial septal defect (ASD) or ventricular septal defect (VSD).

Symptoms

Severe Cyanosis at Birth

Severed Cyan-crayon with Newborn

The parallel circulatory system results in deficient oxygen supply to the tissues, causing severe cyanosis after birth. Cyanosis is the bluish skin tone caused by the presence of deoxygenated hemoglobin. The severity of the cyanosis is based on the amount of mixing between the two parallel circulations. The presence of a shunt, such as PDA, VSD or patent foramen ovale will improve intracardiac mixing and may decrease the amount of cyanosis.

Single S2 sound

Single Stethoscope girl in a (2) Tutu

A second heart sound is often single with increased intensity.

Diagnosis

Echocardiogram

Echoing Cardiogram

Postnatal diagnosis is typically made via echocardiogram. It is important to make the diagnosis as quickly as possible so that treatment can begin immediately.

Chest X-ray shows "Egg-shaped Silhouette"

Egg on Chest X-ray

A chest radiograph may show an "egg shaped silhouette" or "egg on a string" appearance.

Treatment

Prostaglandin

[P-rasta](#)

Initial treatment consists of keeping the ductus arteriosus open with IV PGE1 infusion. This allows some mixing of the two blood circulations.

Avoid NSAIDs

[Avoid-sign N-sad](#)

NSAIDs such as indomethacin close the ductus arteriosus, so it is crucial that NSAIDs are not given to these patients.

Surgical Correction

[Surgeon with scalpel](#)

The arterial switch operation is the most common procedure that permanently corrects the anatomic defect by switching the vessels to their usual positions — the pulmonary artery arising from the right ventricle and aorta from the left ventricle. Coronary arteries (small arteries that supply blood to the heart muscle) must also be moved and reattached to the aorta.

Balloon Atrial Septostomy

[Balloon with Atrial Opening](#)

Balloon Atrial Septostomy is performed to stabilize patients with severe hypoxemia. In this procedure, a balloon is placed across the atrial septum allowing for intracardiac mixing, resulting in improved systemic oxygenation.