Persistent Truncus Arteriosus

Truncus Arteriosus is one of the 5 T's of cyanotic heart defects, along with Transposition of the Great Vessels, Tricuspid Atresia, Tetralogy of Fallot, and Total Anomalous Pulmonary Venous Return. Also known as Common Arterial Trunk, this is a condition in which a single truncal artery gives rise to both the aorta and pulmonary arteries. The cause is unknown, however it has been linked to maternal factors such as advanced maternal age and cigarette smoking, as well as genetic factors. It generally presents with difficulty breathing and poor feeding in the neonate due to pulmonary congestion, leading to heart failure if left unrepaired. Truncus Arteriosus is diagnosed by echocardiogram, and the only definitive treatment is early surgical repair to avoid the development of pulmonary hypertension.



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Common Arterial Trunk
One Common Arterial Trunk
During organogenesis, a single truncal root is divided into the aorta and the pulmonary arteries by the truncoconal septum.

Single Trunk Emerges from Both Ventricles
Single Tree-trunk Emerges from Both V-ventricles
Failure of this process results in a single aortic trunk with a single semilunar valve arising from both ventricles. This causes mixing of systemic and pulmonic venous blood, as the output of both ventricles enters the common trunk.

Symptoms

Cyanosis
Cyan-crayon
Cyanosis is a bluish discoloration of the skin that is caused by abnormally high amounts of deoxygenated hemoglobin in the blood. Cyanosis may not be detectable on physical exam until oxygen saturations drop below 80%, but will be detectable using pulse oximetry. In other instances, neonates may be cyanotic with severe dyspnea.

Pulmonary Congestion
Lungs with Congested-traffic
Pulmonary blood flow increases as arterial resistance naturally falls. This manifests in the infant as difficulty breathing, poor feeding, lethargy, and tachycardia. This also puts the infant at a higher risk of respiratory infections.

Heart Failure
Dead Heart
If left unrepaired, the increased work of pumping blood against the high pulmonary vascular resistance leads to heart failure.

Single S2 Sound
Single Stethoscope-girl in a (2) Tutu
Because there is only one valve, there is only a single, loud second heart sound. There may also be an ejection click heard at the apex or left sternal border. Bounding pulses are also present due to the widened pulse pressure.

Diagnosis

Echocardiogram
Echoing Cardiogram
Diagnosis is made with echocardiography when there is visualization of a single, overriding vessel arising from the heart. Echo also gives information on the truncal valve and root anatomy, and degree of regurgitation.

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Chest X-ray

Chest X-ray

Chest radiographs typically show an enlarged cardiac silhouette due to right and left ventricular hypertrophy. Prominent pulmonary vascular markings may also be visible due to increased blood flow in the pulmonary vasculature.

Treatment

Early Surgical Correction

Early-sun with Surgeon

Preparations for surgical repair should be started as soon as the diagnosis of truncus arteriosus is made. This involves stabilization of cardiopulmonary function supportive care. There is an improved survival rate when primary surgical correction is completed within the first month of life. After repair, some patients will require life-long endocarditis prophylaxis with antibiotics.

Avoids Pulmonary Hypertension

Avoid-sign at Lungs with Hiker-BP

Surgical repair must be completed early to avoid the irreversible development of pulmonary hypertension.

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