Total Anomalous Pulmonary Venous Return (TAPVR)

Total anomalous pulmonary venous return (TAPVR) occurs when the left atrium fails to form a connection with the pulmonary venous plexus in the first month of life, resulting in persistence of the primitive drainage pattern which bypasses the left atrium and drains oxygenated blood into the systemic venous system. This causes cyanosis, fluid overload, and right heart strain. TAPVR can be obstructive or nonobstructive, which makes the severity of symptoms vary greatly. Regardless of degree of obstruction, surgery is the definitive treatment for all levels of TAPVR.



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Pathophysiology

Pulmonary Veins Do Not Connect to Left Atrium

Lung Vine-vein Not Connected to Left A-atrium

TAPVR occurs when the left atrium fails to form connection with the pulmonary venous plexus in embryogenesis, but instead retains its primitive vascular connections. There are several variants, which depend upon the connections retained. In all variants of TAPVR, the pulmonary veins have no connection with the left atrium, and instead empty into the systemic venous system.

Oxygenated Blood Return to Right Atrium

O₂ Blood Returning to Right A-atrium

Due to these aberrant connections, oxygenated blood from the lungs is returned to the right atrium and mixes with the deoxygenated blood returning from the systemic circulation. Because the right atrium is receiving blood from both the systemic and pulmonary circulations, both the right atrium and right ventricle become dilated due to fluid overload.

Incompatible with Life without a Shunt

Lifeguard can't save a Life Without Shunting blood

Without a shunt between the right and left sides of the heart, there is no way for oxygenated blood to be circulated to the body, and TAPVR is incompatible with life. Blood is most commonly shunted right-to-left at the atrial level through an atrial septal defect (ASD). It may also be shunted through a ventricular septal defect (VSD) or a patent ductus arteriosus (PDA).

With Obstruction

Vessel Enters at an Acute Angle

Vessel with Acute-angle

Obstruction can be caused by the pulmonary vein coursing between other vessels, or connecting to the systemic system at an acute angle, causing a pressure backup. If the vein courses inferiorly to the heart, it can be compressed at the level of the diaphragm or at their connections to the portal vascular system.

Severe Cyanosis

Severed Cyan-crayon

Patients with severe obstruction present within the first 12 hours of life with severe cyanosis, respiratory failure, and shock. They are tachypneic with respiratory distress, and diminished cardiac output causes hypotension and diminished pulses.

Pulmonary Edema

Lungs Edamame

Elevated pulmonary vascular pressure in obstructed patients causes the accumulation of fluid in the lungs, which will show up on chest x-ray as pulmonary edema.

Without Obstruction

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Delayed Manifestation (1 or 2 years)

Delayed-child Manifesting tape

Unobstructed forms of TAPVR present later in life, in children 1-2 years old. They present with similar (although less severe) findings of hypoxia, tachypnea, poor feeding, and failure to thrive.

Right Heart Failure

Right Dead Heart

High volume of blood returning to the right heart combined with high pulmonary vascular pressures leads to dilation and hypertrophy, and eventually signs of right heart failure. A fixed, split S2 and systolic ejection murmur often present due to right ventricular overload.

Chest X-ray Shows "Snowman"

Chest X-Ray with Snowman

Chest x-ray in children with unobstructed TAPVR will show normal lung parenchyma with prominence of the pulmonary arteries, innominate vein, and/or SVC leading to the classic “Snowman” or “Figure of 8” sign.

Diagnosis

Echocardiogram

Echoing Cardiogram

Echocardiogram is used to identify the abnormal pulmonary venous connections in infants with suspected TAPVR. It may be able to directly identify abnormal vascular connection points, estimate the pressures in the pulmonary arteries, view the dilation of the right heart, and assess the degree of intracardiac shunting. When echo is equivocal, X-ray, CT or MRA can be used to further define the anatomy.

Treatment

Surgical Correction

Surgeon with scalpel

Initial treatment of infants with severe TAPVR is aimed at stabilization for surgery. Patients are given oxygen, and mechanically ventilated if necessary. Inotropic medications are used to increase the contractility of the heart, and prostaglandins may be beneficial in keeping the ductus arteriosus open to increase the systemic cardiac output. A PDA may also worsen cyanosis, so prostaglandins should be used with caution. Surgical management of TAPVR is recommended regardless of the presence or absence of obstruction, and should be performed as soon as the patient is stable. The goal of surgery is to create a direct pathway between the pulmonary veins and the left atrium.

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